Desmoplastic Fibroma of Bone- Atypical Presentation of a Rare Tumour

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AUTHOR'S CONTRIBUTION

Saurabh Dhaka: Author contributed to the collection of Data, writing of the manuscript, conducting literature review and editing the final case report
Swati Gupta: Author contributed to the reviewing the manuscript, critically revising it for important intellectual content and editing the final case report.
Anju Garg: Author contributed to the reviewing the manuscript, critically revising it for important intellectual content and editing the final case report.

DISCLOSURES

None

CONSENT

Informed oral consent was obtained from the patient prior to submission of the manuscript.

ETHICS AND HUMAN RIGHTS STATEMENT

This case report is based on a patient included in my post graduate thesis, which was approved by the institutional Ethics committee of Maulana Azad Medical college (Approval number: F.1/IEC/MAMC/MD/MS (92/04/2022/No. 177). Informed consent was obtained from the patient at the time of data collection as part of thesis protocol.

ABSTRACT

Desmoplastic fibroma of bone is a rare locally aggressive benign bone tumor. We describe the case of a 38-year-old female patient who presented with gradually progressive swelling of the right proximal arm for 3 years. The radiograph and computed tomogram revealed multiple thin linear and curvilinear radiopacities extending from the cortical surface of the proximal humerus, geographic bone destruction, and cortical disruption. On magnetic resonance imaging, the lesion was replacing the proximal medullary cavity with an associated large extraosseous soft tissue component demonstrating heterogeneous iso to hypointense signal on T2 weighted images. The biopsy revealed haphazardly arranged spindle cells in the background of dense collagenous stroma suggestive of desmoplastic fibroma. The patient underwent wide resection followed by reconstruction with mega prosthesis. The spectrum of imaging characteristics on radiograph, computed tomography, and magnetic resonance imaging along with the management are discussed in detail. Radiologists need to identify typical imaging characteristics of desmoplastic fibroma, which can mimic aggressive malignant bone tumors radiologically. Its locally aggressive behavior and high recurrence rate compared to other benign lesions necessitates a wide excision.

CASE REPORT

Basic information

A 38-year-old female patient presented to the outpatient department with the complaint of gradually progressive long-standing swelling in the anterolateral aspect of the right proximal arm for 3 years. It was associated with recent onset pain from the last 15 days, described as continuous dull aching which is partially relieved with rest and painkillers. There was no history of fever, weight loss, or loss of appetite. On clinical examination, an oval swelling, hard in consistency with ill-defined margins was seen. The overlying veins were prominent and restriction of movement at the glenohumeral joint was present. There was no distal neurovascular deficit. Laboratory parameters and past history were insignificant.

Anteroposterior and lateral radiographs (Figure 1) showed a large, ill-defined, oval-shaped, osteolytic lesion epicentered in the metaphysis of the right proximal humerus, extending into adjacent diaphysis and epiphysis reaching up to the subchondral region. The lesion was centrally located within the medullary cavity with a wide zone of transition. The erratic destruction of bone was evident as irregular coarsened bony ridges traversing within the lesion representing the characteristic internal pseudo trabeculation. Aggressive disorganized periosteal reaction

was seen as multiple linear and curvilinear reticular strands extending from the cortical surface. The associated endosteal scalloping and aggregation of periosteal new bone formation resulted in irregular cortical thickening and mild expansile remodeling of bone. A pathological fracture and loss of cortical integrity were seen with the resultant extension of the lesion into the extraosseous soft tissue. The extraosseous soft tissue had relatively preserved fascial planes with adjacent muscles. These radiographic features suggested an atypical aggressive osteolytic lesion and differential diagnosis of chondrosarcoma or osteosarcoma were kept. For further characterization of the lesion, non-contrast computed tomography (NCCT) and contrast-enhanced magnetic resonance imaging (CE-MRI) were performed.

CT images (Figure 2) confirmed the radiographic findings of an ill-defined intramedullary lesion with geographic areas of bone destruction with few internal pseudo trabeculae. Cortical break, irregular cortical thickening, endosteal scalloping, and disorganized periosteal reaction are well demonstrated on bone window images of NCCT. No distinctive matrix mineralization was seen.

MRI (Figure 3) of the right humerus showed replacement of intramedullary fat by the lesion measuring ~13.4 (CC) x 10.3(TR)x 8.2(AP)cm. The signal intensity of the lesion was slightly hyperintense to muscle on T1 weighted images (Figure 3A) and, heterogeneous hyperintense signal on T2 weighted images (Figures 3B,3C) with multiple hypointense areas. Both intraosseous and extraosseous components showed heterogeneous enhancement (Figure 3D) following intravenous contrast administration. The lesion was extending along the medullary cavity up to the articular cortex, however, joint invasion was absent. The extraosseous component had multilobulated well-defined margins and was causing displacement of adjacent muscles. The adjacent brachial neurovascular bundle and radial nerve (in the radial groove of the humerus) were in proximity to the lesion but fat planes were maintained.

To reach a definitive diagnosis, open biopsy was done along the lateral aspect through the deltoid muscle causing minimal interference with fascial planes to avoid local spread of tumor cells. On examination, biopsy tissue revealed woven and lamellar bone fragments with the replacement of intertrabecular spaces by haphazardly arranged bland spindle-shaped cells in dense fibro collagenous tissue of low cellularity (Figure 4B). No evidence of brisk mitosis or pleomorphism was seen. The spindle-shaped cells were positive for myoepithelial differentiation marker SMA (Figure 4D), focal cytoplasmic positivity for beta-catenin (Figure 4D), and nucleus diffusely stained for expression of H3K27me3. They were negative for desmin, S-100, and H-caldesman. These findings are diagnostic of benign desmoplastic fibroma of bone.

The patients underwent en-bloc excision of the tumor. A wide resection was done and a segment of 18 cm of the proximal humerus was resected including adequate safe margins of 5cm to reduce the risk of recurrence. Resection was followed by reconstruction with a mega prosthesis. The postoperative period was uneventful and no neurological or vascular deficit was observed. Gross examination (Figure 4A) of the resected specimen revealed an expansile lesion covered by a thin capsule and no areas of capsular breach were identified. On the cut section, the tumor was yellowish in color and hard in consistency. On pathological examination, margins were clear of tumor and the lesion was partially capsulated with focal areas of capsular breach and invasion of tumor into adjacent skeletal muscles. Histopathological examination of the specimen revealed features similar to preoperative biopsy.

The patient was followed up at one month, 3 months, 6 months, and 10 months with radiographs. The patient resumed her day-to-day activities after 3-4 months of surgery, however slight restriction in overhead abduction was present. In the last follow-up at the duration of 10 months, no fresh complaints were observed and no clinical or radiographic evidence of recurrence was present.

DISCUSSION

Etiology and demographics

Desmoplastic fibroma is a rare bone tumor comprising less than 0.1% of primary bone tumors [1]. It was first described by Jaffe in 1958 [2]. The World Health Organization (WHO) classification of bone tumors has categorized desmoplastic fibroma as a fibrogenic tumor of bone of intermediate grade [3]. It can occur in any age group but most commonly adolescents and young adults are affected with peak incidence in the 2nd and 3rd decade of life and no gender predilection are seen [4]. Desmoplastic fibroma of bone is a slowly growing locally aggressive tumor that does not metastasize, and patients commonly present with complaints of dull aching pain and swelling for long durations. Rarely it can be incidentally detected on imaging. The occurrence of desmoplastic fibroma is equally likely to involve the axial or appendicular skeleton involving both flat and long bones, the most commonly affected bone is the mandible followed by the femur [3,5]. On histopathology, the tumors show a resemblance with their non-osseous counterpart, desmoid fibroma.

Imaging findings

The radiographic appearance of desmoplastic fibroma has a resemblance similar to other benign lytic bone lesions, however, a few specific features can help to differentiate it from other lesions. Typically, desmoplastic fibroma involves the metaphysis of long bones and can extend into epiphysis and diaphysis. It has well-defined margins with a narrow zone of transition and causes geographic lytic destruction with non-sclerotic margins. Internal pseudo trabeculation is the characteristic feature of fibrogenic tumors, formed due to non-uniform destruction of bone. Expansile remodeling of bone suggests the continuous benign slow-growing nature of the

lesion. Pathological fractures can be seen in 10-15 % of cases [6,10]. Features of aggressiveness can be seen in the form of cortical disruptions and extraosseous extension of lesions. Cortical destruction is seen in up to 29% of the patients [6,8]. It is the consequence of slow but prolonged osseous destruction and should not be confused as a sign of malignancy. However, invasion in adjacent muscles is rarely seen, rather the tumor displaces the adjacent muscles. Periosteal reaction and matrix mineralization are unusual phenomena in desmoplastic fibroma [6]. These radiographic findings represent a benign lesion showing a slow growth rate with focal areas of aggressive behavior. CT demonstrates the bone details of the lytic lesion accurately and the radiographic findings are corroborated.

However, our case had an unusual appearance showing features of aggressive malignant bone lesions such as a wide zone of transition, ill-defined margins, disorganized aggressive periosteal reaction, cortical destruction, pathological fracture, and extraosseous extension. These features directed us toward a differential diagnosis of more aggressive malignant bone lesions.

There is a paucity of literature describing features of desmoplastic fibroma on MRI. Like any other bone tumor, it appears as an altered area of signal intensity involving the medullary cavity. However, a characteristic feature of desmoplastic fibroma that narrows down the diagnosis is the presence of areas of low signal intensity compared to skeletal muscles on T2W images that were inconsistent with the areas of calcification seen on corresponding NCCT images. These low signal areas occur due to T2 shortening by the presence of a dense fibro-collagenous matrix [7,8]. Although, the lesion has predominantly high signal areas on T2W images represents areas of high cellularity, loose collagen arrangement, and cystic degeneration or necrotic areas. Similar T2 hypointense signal intensity is seen in fibromatosis, GCT, lymphoma, and fibrous dysplasia. It appears iso to hypointense to adjacent skeletal muscles on T1 W images [8,9]. The lesion shows heterogenous enhancement on post-contrast sequences depicting the diversified matrix composition- collagen, myofibroblast, necrotic areas, and remaining trabecular bone. The areas showing intense enhancement correspond to areas of high cellularity and the remaining areas with minimal or no enhancement can be described as areas of dense collagen and bony fragments.

Histopathological examination

The histopathological distinction between desmoplastic fibroma of bone and its soft tissue counterpart desmoid tumor is an arduous task, due to the overlap of the majority of findings. Though accumulation of beta-catenin in the nucleus is diagnostic of desmoid fibroma, this is not commonly seen in the case of desmoplastic fibroma. The classical histopathological appearance of desmoplastic fibroma is made of woven and lamellar bone fragments with the replacement of intertrabecular spaces by haphazardly arranged fascicles of bland spindle cells, fibroblast and myofibroblast cells in dense fibro collagenous matrix of low cellularity. Features suggestive of malignancy such as nuclear atypia, brisk mitosis, pleomorphism, and coarse chromatin are scarcely visualized, few giant cells can be seen due to the chronic inflammatory process. On immunohistochemical staining, the majority of tumor cells are positive for cytoplasmic SMA [11] and negative for desmin, S100, and keratin. In our case histological appearance was in concordance with other studies, the tumor cells were positive for SMA and cytoplasmic beta-catenin and negative for nuclear beta-catenin.

Management and prognosis

Desmoplastic fibroma can be managed by several treatment strategies such as surgery, radiotherapy, and pharmacotherapy. However, a major issue that needs to be encountered in its management is its high local recurrence rate. Surgery is the preferred treatment modality due to the lowest recurrence rates. Various surgeries defined in the literature include curettage, marginal excision, and wide resection. Recurrence rates following curettage were up to 42% -55% [10, 12], and were lowest in wide resection (up to 17 %). In our case, wide resection was performed followed by reconstruction with mega prosthesis. No clinical or radiographic signs of recurrence were observed till 10 months of follow-up.

Differential diagnosis

Various lesions can mimic desmoplastic fibroma based on these radiographic features and a radiologist needs to differentiate desmoplastic fibroma from giant cell tumor (GCT), chondromyxoid fibroma, telangiectatic osteosarcoma, aneurysmal bone cyst (ABC) as well as fibrosarcoma. When features such as wide transition zone, cortical destruction with soft tissue extension, and aggressive periosteal reaction are seen, the appearance of desmoplastic fibroma can imitate malignant bone lesions like osteosarcoma, chondrosarcoma, and fibrosarcoma.

Chondromyxoid fibroma

Chondromyxoid fibroma can occur in any age group, the majority of the cases are below 30 years of age. On the radiograph, it appears as well well-defined eccentric expansile lytic lesion with internal septations involving metaphysis of long bones. The pseudo trabeculation seen in chondromyxoid fibroma is curvilinear to a greater extent compared to desmoplastic fibroma. Periosteal reaction and matrix mineralization are commonly not seen. However, it can be distinguished from desmoplastic fibroma by their eminent homogenous hyperintense signal on T2 weighted imaging.

Aneurysmal bone cyst (ABC)

ABC commonly occurs in the first two decades of life and appears as well defined expansile lytic lesion with a soap bubble appearance in the metaphysis of long bones on the radiograph. It can be distinguished by its characteristic fluid-fluid levels on MRI. Cortical break and periosteal reactions are rarely seen in ABC [13].

Giant cell tumor of bone

Giant cell tumors occur distinctively in the subchondral region of epiphysis in skeletally mature individuals. On the radiograph, it appears as an eccentric expansile lytic lesion, subchondral in location giving a soap bubble appearance. It causes geographic lytic destruction with well-defined margins. Periosteal reaction and matric mineralization are not a common feature. On histopathological examination, multiple giant cells in the background of mononuclear cells can confirm the diagnosis.

Telangiectatic osteosarcoma

It is an aggressive osteogenic bone tumor, mainly composed of multiple variable-sized hemorrhagic cavities. Patients typically present with a short history of gradually progressive pain and swelling. On radiograph, an ill-defined lesion with a wide zone of transition is seen involving the metaphysis of long bones. There is an aggressive type of periosteal reaction and permeative bony destruction. Cortical breaks and extraosseous extensions are frequently seen. The matrix can show areas of osteoid mineralization. On MRI, blood-fluid and fluid-fluid levels preoccupy the majority of tumor volume along with an enhancing solid component that differentiates it from other lesions showing fluid-fluid levels.

TEACHING POINT

Desmoplastic fibroma of bone is a locally aggressive nonmetastatic bone tumor that appears as a well-defined expansile lesion involving the metaphysis with geographic bone destruction and internal pseudo-trabeculations on plain radiographs. MRI shows a marrow-replacing lesion with hypointense signal on T2W images. Biopsy is required to make a definitive diagnosis which reveals spindle-shaped cells positive for SMA in a dense fibrocollagenous matrix. Wide excision of the lesion should be done to minimize the risk of recurrence. This case reports an atypical presentation of this rare bone tumor, however a radiologist should identify the characteristic features on radiograph and MRI to narrow down the differential diagnosis and differentiate it from aggressive malignant bone tumors for appropriate timely management.

QUESTIONS

Question 1: A 32-year-old female presented with a complaint of pain and swelling in the distal femur for the last 3 years, on clinical examination a firm swelling with tenderness was noted. The radiograph showed a well-defined expansile lytic lesion involving metaphysis with internal trabeculations, geographic lytic bone destruction with nonsclerotic margins, and no matrix mineralization or periosteal reaction. Based on these findings, which of the following is the most likely diagnosis?

A) Osteosarcoma
B) GCT
C) Desmoplastic fibroma [applies]
D) Bone metastasis
E) Chondrosarcoma

ANS: The correct option is C) Desmoplastic fibroma- occurs in young adults with the peak incidence in the 2nd and 3rd decade. Benign lesion occurring in the metaphysis of long bone. On the radiograph appears as well a well-defined lytic lesion with nonsclerotic margins and internal pseudo-trabeculation.[It can occur in any age group but most commonly adolescents and young adults are affected with peak incidence in the 2nd and 3rd decade of life and no gender predilection is seen]

Incorrect options

Osteosarcoma: shows a bimodal distribution, before 20 years, and after 60 years, osteoid matric mineralization and aggressive periosteal reaction is seen.

GCT occurs in the epiphysis of long bones.

D)Bone metastasis seen in older age group. Presenting history is of short duration. **E)Chondrosarcoma-** malignant lesion with predilection for flat bones. Radiographic features include aggressive periosteal reaction and ring and arc type of matrix mineralization.

Question 2: Which of the following is the most appropriate treatment for a patient with a desmoplastic fibroma of the distal femur?

A) Radiotherapy
B) Wide resection [applies]
C) Curettage
D) Chemotherapy
E) Amputation

Answer: The correct answer is option B- wide resection of the lesion, desmoplastic fibroma has a high local recurrence rate so wide resection is preferred management. [Recurrence rates following curettage were up to 42% -55% [10, 12], and were lowest in wide resection (up to 17 %)]

Incorrect options: A) Radiotherapy has a very limited role in the management of desmoplastic fibroma as tumor cells do not show any brisk mitosis or pleomorphism.

C) Curettage has a high recurrence rate so not preferred.

D) chemotherapy has no role as tumor cells do not show any brisk mitosis or pleomorphism.

E) Amputation is done for high-grade malignant bone tumors in which limb salvation cannot be done.

Question 3: Which of the following best describes the appearance of desmoplastic fibroma on MRI?

A) Lesion replacing medullary fat, appearing hyperintense on T1 images, hyperintense on T2 images, and does not show any post-contrast enhancement.

B) Lesion replacing medullary, fat appearing hypointense on T1 images, hyperintense on T2 images, and show homogeneous post-contrast enhancement.

C) A cortical-based lesion with normal medullary fat, appears hypointense on T1 images, hyperintense on T2 images, and shows homogeneous post-contrast enhancement.

D) Lesion replacing medullary fat appearing hypointense on T1 images, hyperintense on T2 images with hypointense areas and show heterogeneous post-contrast enhancement. [applies]

E) A cortical-based lesion with normal medullary fat appearing hyperintense on T1 images, hyperintense on T2

images with hypointense areas, and shows heterogeneous post-contrast enhancement.

Answer: the correct answer option is D) Desmoplastic fibroma appears as a well-defined lesion replacing medullary fat appearing hypointense on T1 images, hyperintense on T2 images with hypointense areas, and show heterogeneous post-contrast enhancement. It is a fibrogenic tumor with a dense fibro collagenous matrix responsible for T2 hypointense areas and heterogeneous post-contrast enhancement.[These low signal areas occur due to T2 shortening by the presence of a dense fibro-collagenous matrix. Although, the lesion has predominantly high signal areas on T2W images represents areas of high cellularity, loose collagen arrangement, and cystic degeneration or necrotic areas]

Question 4: Which of the following best describes desmoplastic fibroma of bone?

A) WHO high-grade tumor of fibro-genic origin with a high propensity for metastasis.

B) WHO intermediate-grade tumor of fibro-genic origin with a high propensity for metastasis.

C) WHO low-grade tumor of fibro-genic origin which does not show how metastasis.

D) WHO intermediate-grade tumor of fibro-genic origin which does not show metastasis.[applies]

E) WHO low-grade tumor of fibro-genic origin with a high propensity for metastasis.

Answer: the correct answer is option D) WHO intermediate-grade tumor of fibro-genic origin which does not show metastasis. Desmoplastic fibroma is categorized as WHO intermediate grade, a locally aggressive tumor of fibrogenic origin that does not show metastasis. [The World Health Organization (WHO) classification of bone tumors has categorized desmoplastic fibroma as a fibrogenic tumor of bone of intermediate grade]

Question 5: which of the following best describes the histopathological findings of desmoplastic fibroma?

A) Spindle-shaped cells in the background of dense fibro-collagenous matrix. Tumor cells positive SMA and cytoplasmic beta-catenin.[applies]

B) Spindle cells with nuclear atypia, show brisk mitosis and pleomorphism.

C) Multinucleate giant cells in the background of mononuclear cells.

D) Tumor cells with marked anaplasia and pleomorphism showing bone formation with interspersed osteoclastic cells.

E) Multiple round cells with high Nuclear: cytoplasm ratio and positive for vimentin, CD 99, and NSE.

Answer: the correct option is A) Spindle-shaped cells in the background of dense fibro-collagenous matrix. Tumor cells positive SMA and cytoplasmic beta-catenin. No nuclear atypia or pleomorphism is seen.[haphazardly arranged fascicles of bland spindle cells, fibroblast and myofibroblast cells in dense fibro collagenous matrix of low cellularity]

Incorrect options

B) In desmoplastic fibroma, nuclear atypia or pleomorphism are not seen.

C) Multinucleate giant cells in the background of mononuclear cells are seen in GCT.

D) Tumor cells with marked anaplasia and pleomorphism showing bone formation with interspersed osteoclastic cells- this histological appearance is seen in osteosarcoma.

E) Multiple round cells with high Nuclear: cytoplasm ratio and positive for vimentin, CD 99, and NSE these features represent Ewings sarcoma.

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FIGURES

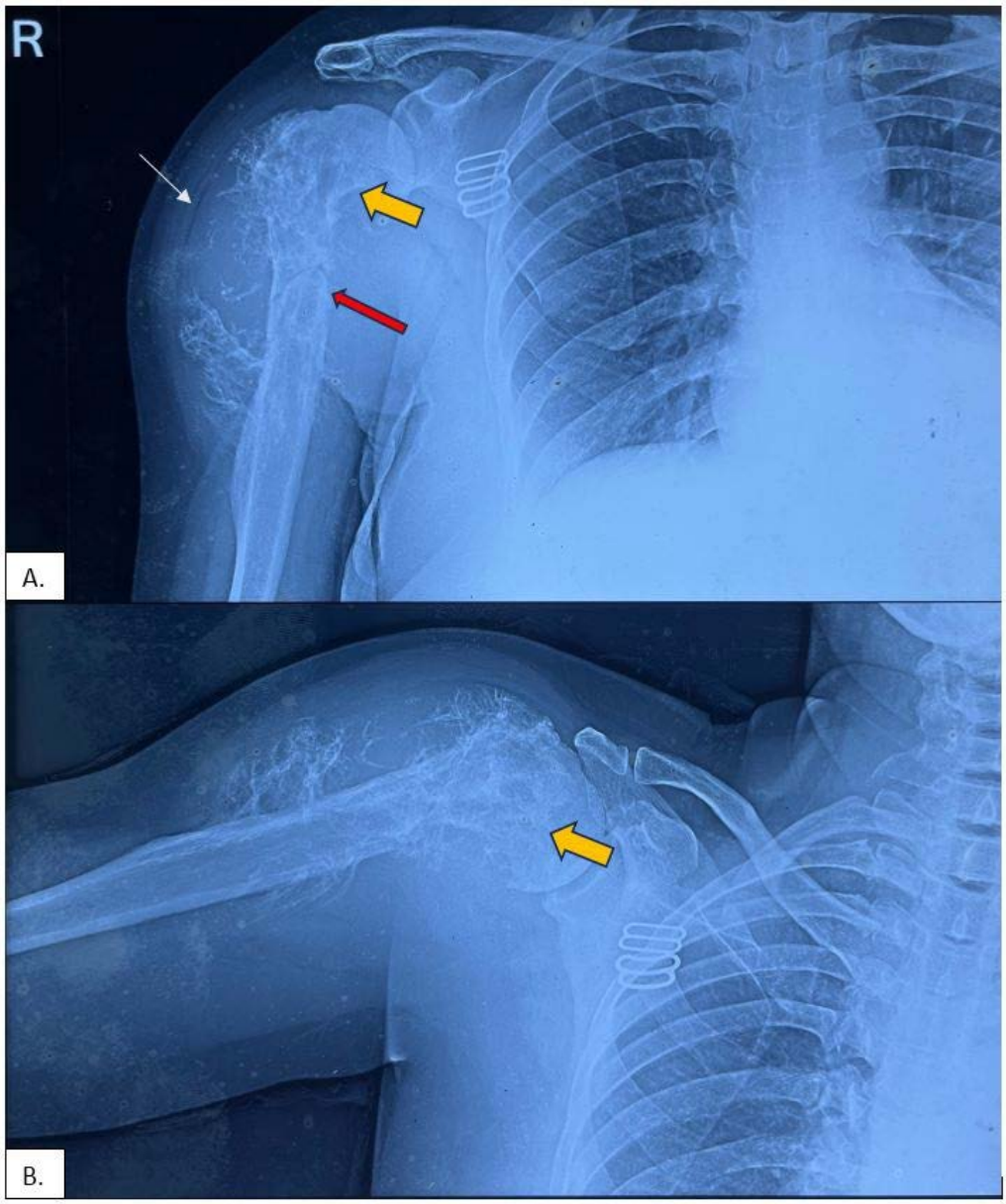

Figure 1: 38-year-old female with desmoplastic fibroma in the right proximal humerus.

FINDINGS: AP radiograph of the right shoulder in adduction and abduction demonstrates an ill-defined lesion with a wide transition zone showing internal pseudo-trabeculations (yellow arrow), disorganized periosteal reaction, and cortical breakthrough. The extraosseous component is displacing the adjacent muscles evident as maintained fascial planes (white arrow). Pathological fracture is also seen along the medial cortex (red arrow).

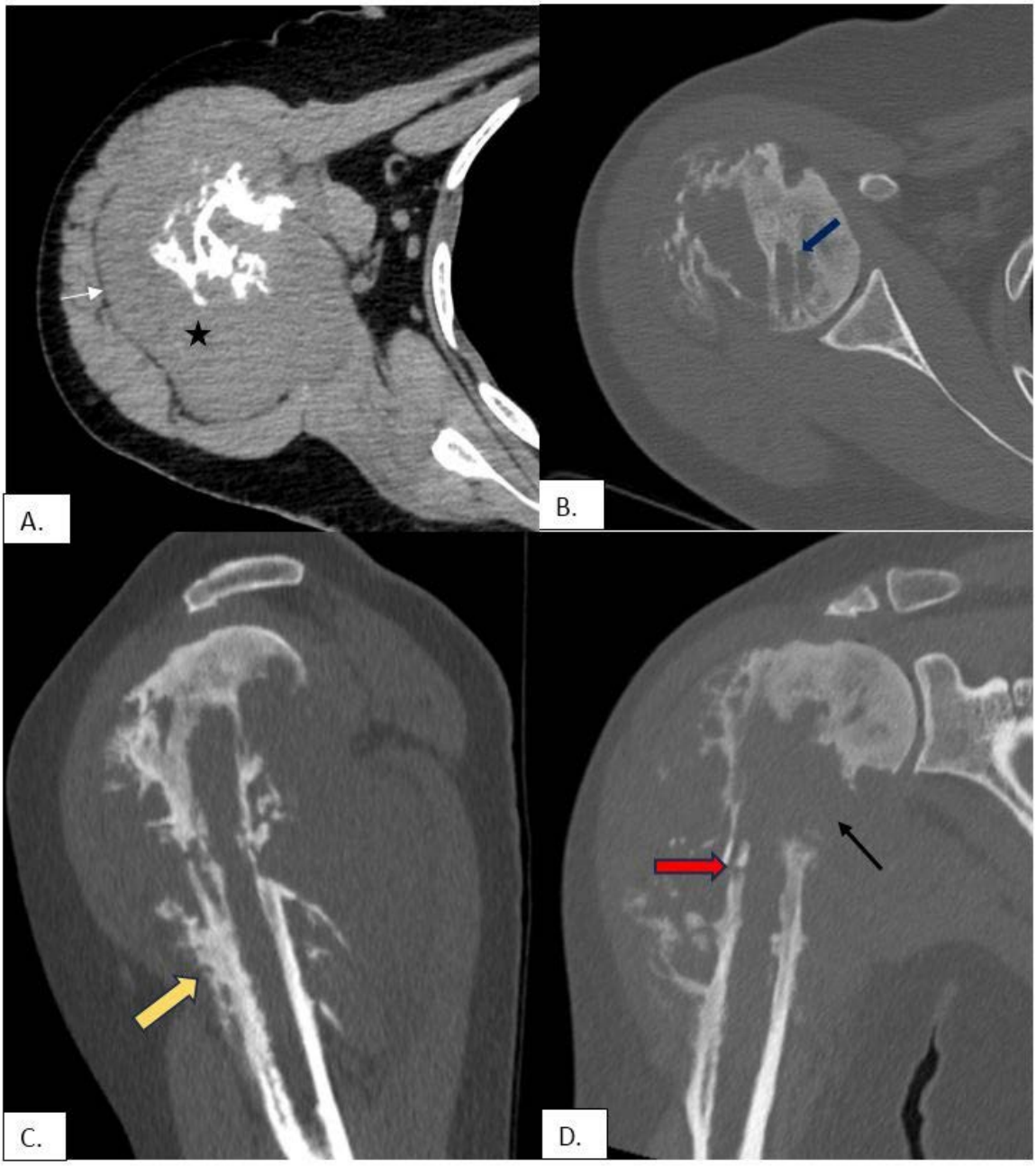

Figure 2: 38-year-old female with desmoplastic fibroma in the right proximal humerus.

TECHNIQUE:
Noncontrast computed tomography images of the right shoulder with proximal humerus were obtained on a GE 128 slice scanner with settings of kVp 140, mA 400, and slice thickness 0.625mm in helical axial acquisition.
FINDINGS:
(A) Axial soft tissue window reformats demonstrate an ill-defined oval-shaped lytic lesion with an extra-osseous soft tissue component (star). The lesion is predominantly displacing adjacent muscles with relatively maintained fascial planes (white arrow).
(B) Axial bone window reformat shows non-uniform destruction of bone which results in internal pseudo-trabeculation.
(C) Sagittal bone window reformats demonstrate aggressive disorganized periosteal reaction with cortical irregularities.
Coronal bone window reformats demonstrate areas of cortical destruction (black arrow) and pathological fracture (red arrow).

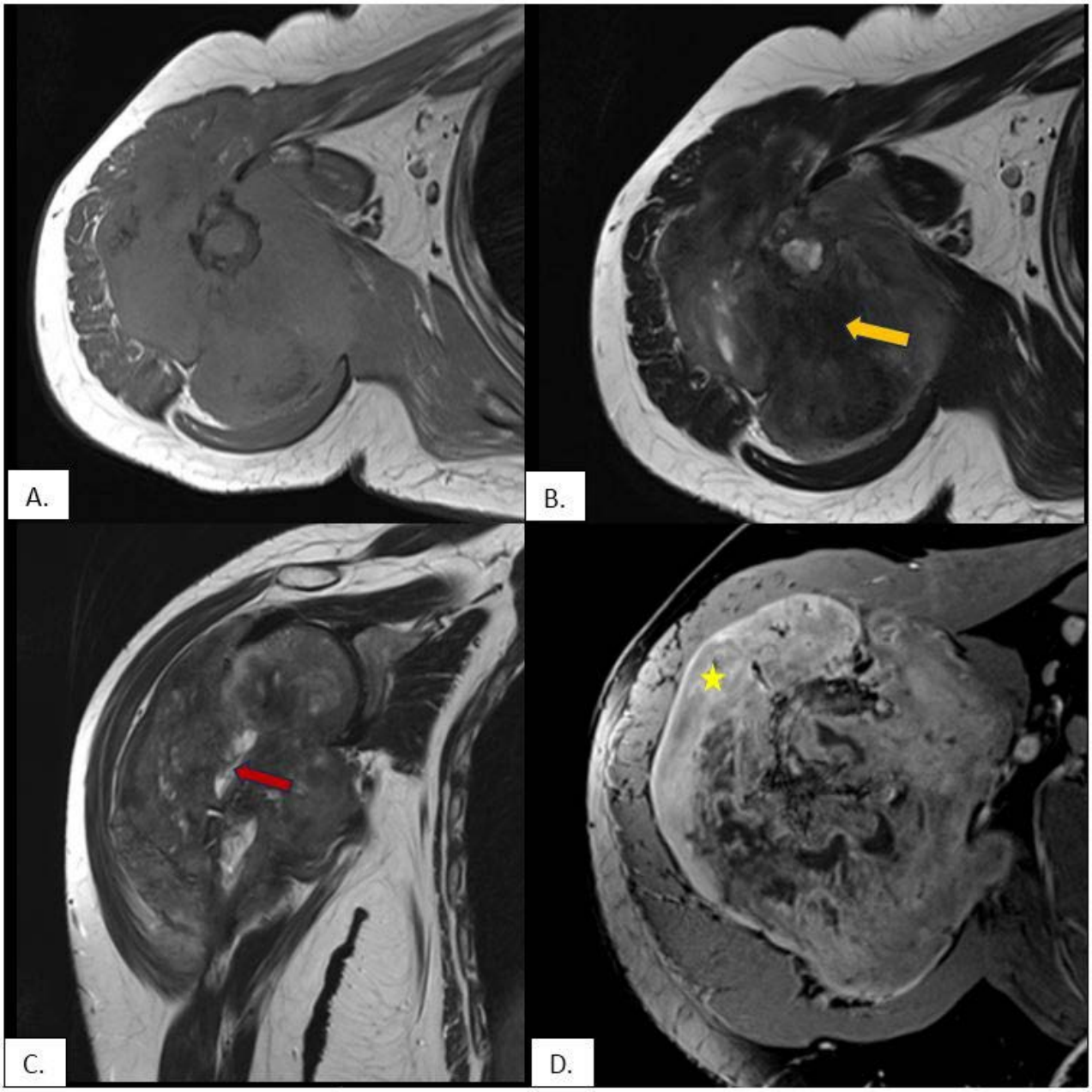

Figure 3: 38-year-old female with desmoplastic fibroma in the right proximal humerus.

FINDINGS: Pre-contrast and post-contrast MR sequences were acquired from the right shoulder joint
(A) Axial T1-weighted image demonstrating a large ill-defined lesion replacing marrow that is isointense to hyperintense to muscle.
(B) Axial and (C) coronal T2-weighted images show a large ill-defined lesion with heterogeneous signal intensity with internal low-intensity areas depicting dense fibro collagenous matrix, the high signal areas represent areas of high cellularity and few cystic and necrotic areas (red arrow in C).
(D) Post gadolinium FS VIBE image demonstrates heterogeneous post-contrast enhancement of the lesion, areas showing intense enhancement (yellow star) correspond to areas of high cellularity.
TECHNIQUE:
(A) Pre-contrast axial T1 weighted MR image of right shoulder acquired on a 3 Tesla magnet with 4 mm slice thickness (TE-11ms, TR-705 ms).
(B) Pre-contrast axial T2 weighted images of the right shoulder were acquired on a 3 Tesla magnet with 4mm slice thickness (TE-105 ms, TR-3500 ms).
(C) Pre-contrast coronal T2 weighted images of the right shoulder were acquired on a 3 Tesla magnet with 3mm slice thickness (TE- 103ms, TR-4280ms).
(D) Post gadolinium fat-suppressed axial VIBE images of the right shoulder was acquired on a 3 Tesla magnet with a 1mm slice thickness (TE-4.43ms, TR-9.88ms). (0.1 mmol/kg body weight of intravenous Gadopentetate dimeglumine was injected with a power injector at an injection rate of 3 mL/sec followed by a bolus of 20 mL saline solution)

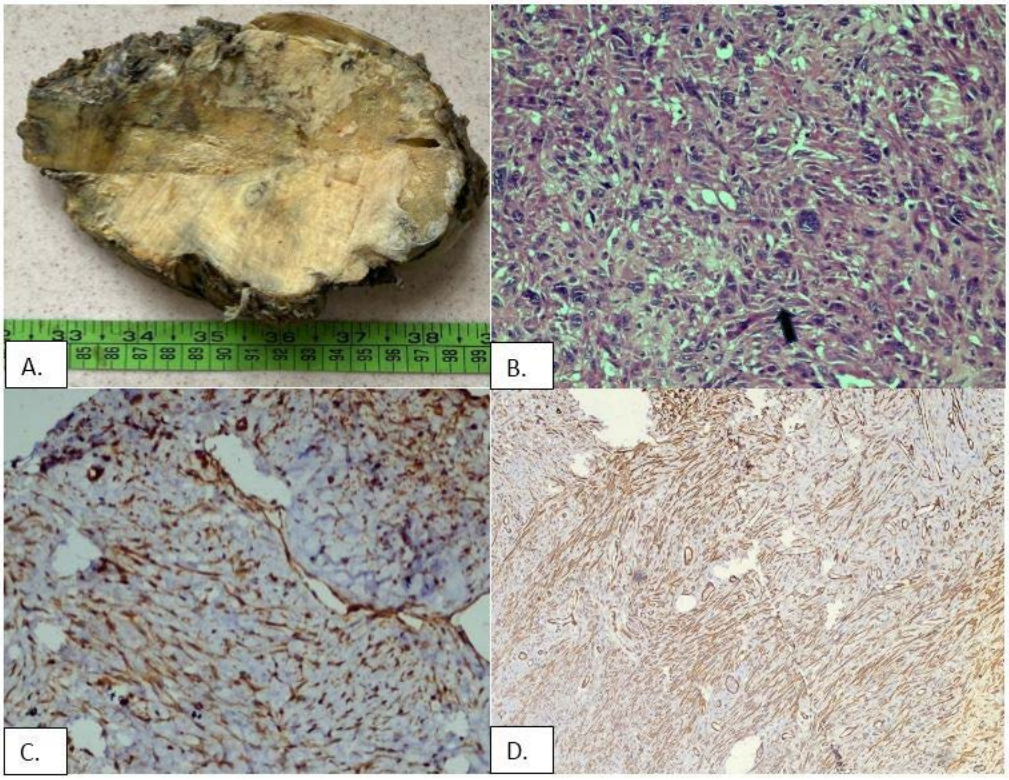

Figure 4: 38-year-old female with desmoplastic fibroma in the right proximal humerus.

FINDINGS:

(A) A gross excision specimen of lesion measuring ~15 x10 x 7cm (CC x TR x AP) appears yellowish with firm to hard consistency. The lesion is covered by a thin capsule with no obvious breach.

(B) Photomicrograph of H& E stained decalcified specimen in 20 x magnification shows a low cellularity tumor with haphazardly arranged fascicles of spindle cells in the background of dense fibro-collagenous matrix.

(C) Immunohistochemical staining, tumor cells stained positive for cytoplasmic Beta-catenin (200x magnification).

Immunohistochemical staining, tumor cells stained positive for SMA.

Table 1: Summary table of desmoplastic fibroma of bone

Etiology	Intermediate-grade fibrogenic bone tumor with locally aggressive behavior
Incidence	Rare bone tumors representing <0.1 % of all bone tumors.
Sex predilection	None
Age predilection	In young adults with peak incidence in 2nd and 3rd decade of life.
Risk factors	No risk factors identified
Finding on imaging	Radiograph and CT: geographic lytic lesion involving metaphysis with internal pseudo trabeculae, cortical irregularity, and destruction with no matrix mineralization and periosteal reaction. MRI: marrow replacing lesion showing both hypointense and hyperintense signal on T2W images and heterogeneous post-contrast enhancement.
Biopsy	Haphazardly arranged fascicles of spindle cells in dense fibro collagenous matrix of low cellularity, cells positive for SMA.
Treatment	Wide local excision is the treatment of choice.
Prognosis	High local recurrence rate- 17% for wide resection and 42-55% for curettage.

Table 2: Differential diagnosis table for desmoplastic fibroma of bone

Lesion	Radiograph/CT Finding	MRI Findings
Desmoplastic Fibroma	• Well defined geographic lytic lesion with non-sclerotic margins involving the metaphysis • Internal pseudo trabeculae, cortical irregularity, and destruction • No matrix mineralization and periosteal reaction.	• Iso to hypointense on T1W images • Marrow replacing lesion showing both hypointense and hyperintense signal on T2W images • Heterogeneous post-contrast enhancement.
Chondromyxoid fibroma	• Well-defined eccentric expansile lesion. • Internal pseudo-trabeculations are curvilinear. • Cortical destruction can be seen	• Low T1W signal intensity • Homogenous high signal on T2W images. • Homogenous post-contrast enhancement.
Aneurysmal bone cyst (ABC)	• Well-defined expansile lesion with internal septation and sclerotic margins giving soap bubble appearance in the metaphysis of long bones. • Cortical destruction and periosteal reaction are not seen. • Geographic lytic destruction.	• Variable signal on T1W images. • Fluid-fluid levels are seen on T2 W images. • Heterogeneous post contrast enhancement.
Giant cell tumor (GCT)	• Well-defined eccentric subchondral geographic lytic lesion. • Expansile lesion giving soap bubble appearance, located in the epiphysis of long bones. • Cortical destruction can be seen.	• Iso to hypointense on T1W images. • Variable signal on T2W images. • May show hemorrhagic component or fluid-fluid levels
Telangiectatic osteosarcoma	• An ill-defined lesion with a wide transition zone. • Aggressive type of periosteal reaction. • Cortical destruction and matrix mineralization can be seen.	• Blood-fluid levels or fluid- fluid levels can be seen on T2 W images along with a solid component. • Areas of mineralization appear as low signal areas on all sequences. • Heterogenous enhancement on post-contrast images.

KEYWORDS

Desmoplastic fibroma, bone tumors, musculoskeletal radiology, T2W hypointense lesion, pseudo-trabeculations

ABBREVIATIONS

NCCT=NONCONTRASTCOMPUTEDTOMOGRAPHY

CE-MRI = CONTRAST ENHANCED MAGNETIC RESONANCE IMAGING

GCT = GIANT CELL TUMOR

SMA = SMOOTH MUSCLE ACTIN

WHO = WORLD HEALTH ORGANIZATION

ABC = ANEURYSMAL BONE CYST