Unusual presentation of more common disease/injury

Diaphyseal desmoplastic fibroma of humerus – report of a rare presentation

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Desmoplastic fibroma is a rare locally aggressive benign tumour. It is most frequently reported in mandible, metaphysis and metadiaphysis of long bones. This tumour has been observed infrequently in the diaphysis like the one presented here. An 18-year-old female presented with progressive swelling left arm and dull aching pain for 1 year. She had a bony hard lobular well-defined swelling in the midshaft of humerus, which was not warm but tender. There was abnormal mobility present indicating a fracture. Plain radiograph of the affected arm showed an expansile, well-defined, intraosseous, radiolucent lesion with expansion involving the entire circumference of the humerus. An open biopsy was carried out and the report came out to be desmoplastic fibroma. She underwent wide segmental resection and interlocking nailing and bone grafting. After 6 weeks follow-up, the patient had visible callus and by 12 weeks there was significant union.

BACKGROUND

Desmoplastic fibroma is a rare locally aggressive benign tumour originally described by Jaffe in 1958.¹ It is thought to be the osseous counterpart of the soft tissue desmoid tumour, with an incidence of 0.11%.² Bohm *et al* in a review of literature reported 189 cases and added two additional patients.³ A review of literature since the report by Bohm *et al*, noted an additional 28 patients primarily in the form of case reports.^{4–13} Known to be a lesion from metaphysis, diaphyseal localisation of the tumour is rarely reported.¹⁴ We intend to report an unusual presentation site of this rare tumour successfully treated at our institution.

CASE PRESENTATION

An 18-year-old female presented with history of swelling in the left arm, which was slowly increasing in size for the past one and a half years. There was no history of trauma or fever. She also had dull aching pain for the past 1 year. Pain initially was activity related, but for the past 3 months, she had severe pain even at times of rest. She had a bony hard lobular well-defined swelling in the midshaft of humerus, which was not warm but tender. There was abnormal mobility present indicating a fracture. There was no palpable lymphadenopathy. Skin over the swelling appeared normal and there was no distal neurovascular deficit. Movements of the elbow and shoulder were normal but caused pain at the fracture site.

INVESTIGATIONS

Haemogram and biochemical investigations were normal (figure 1). Plain radiograph of the affected arm was taken and the following findings were noted. There was an expansile, well-defined, intraosseous, radiolucent lesion involving the entire circumference of the humerus. There was marked thinning of the cortex with delicate and diffuse

trabeculations giving a soap bubble appearance. There was cortical discontinuity indicating pathological fracture. After necessary investigations an open biopsy was carried out. There were fragments of fibrous tissue with dense collagenisation, woven bone and dead bony fragments

Figure 1 Preoperative radiograph.

(figure 2). No mitosis or atypia seen and the overall features suggested a fibrous lesion, most likely desmoplastic fibroma (figure 3).

Figure 2 Follow-up at 1 year.

TREATMENT

On exploration there was a spindle expansile lesion in the midshaft, the periosteum was thickened and there was cortical thinning out. The pathological fracture was made out and there were numerous loculations, which had straw-colored fluid and membranous lining. She underwent wide segmental resection and simultaneous interlocking nailing and bone grafting. Her contralateral fibula and iliac crest were used for grafting. The excised specimen was sent for histopathological examination which confirmed the diagnosis. The limb was protected in a plaster of Paris splint for 6 weeks. After 6 weeks follow-up, the patient had visible callus and by 12 weeks there was significant union (figure 4). There was no functional impairment. The patient is in follow-up for the past 4 years.

DISCUSSION

Desmoplastic fibroma is most frequently reported in mandible, metaphysis and metadiaphysis of long bones. This tumour has been observed infrequently in the diaphysis like the one presented here. Only two previous case reports were there on diaphyseal tumours.^{3 14} Most patients were younger than 30 years of age.^{2 10} Pain is the most common presenting complaint.⁵

There is an approximately 10% to 12% rate of pathological fractures at the time of presentation.^{3 5} Radiographs show expansile, radiolucent lesions. These tumours have well-defined margination with a narrow zone of transition, consistent with their slow growth, without any surrounding bone sclerosis. Due to the internal bony septations they have a honeycombed appearance. Also there is endosteal scalloping and consequent cortical thinning; periosteal reaction being rare.^{7 13} Histopathologic examination reveals a sparsely cellular lesion composed of

Figure 3 Histopathological picture.

Figure 4 Lesion composed of fibrous tissue with dense collagenisation, woven bone and dead bony fragments.

uniform spindle shaped fibroblasts between heavy, parallel bands of collagen fibres. The periphery of the lesion may be more cellular, with infiltration into the surrounding tissues. This makes it difficult to differentiate desmoplastic fibroma from low-grade fibrosarcoma. However, the former lacks the degree of cellularity, pleomorphism, mitotic activity and herringbone pattern seen in low-grade fibrosarcoma.^{3 15} The expansile lytic appearance of this lesion on plain radiograph is suggestive of fibrous dysplasia.

On histopathologic examination, fibrous dysplasia is comprised of loosely arranged fibrous tissue with metaplastic, woven bone formation.² This is in contradistinction to desmoplastic fibroma, which is comprised primarily of sparsely cellular areas with dense thick bands of collagen fibers. Any bone formation within this lesion is isolated to the periphery and is reactive in nature or is the residual bony trabeculae. However, the coexistence of desmoplastic fibroma and fibrous dysplasia has been reported.¹⁶ Therefore adequate biopsy specimens from the centre of the lesion to identify areas with pronounced collagen bundles are necessary for the accurate diagnosis of desmoplastic fibroma.

Diaphysis being an infrequent site of the tumour limits the differential diagnosis. Primary lymphoma when present in long bones is seen most frequently in the diaphysis. Lymphoma appears more aggressive than desmoplastic fibroma and is seen as a permeative lesion with adjacent periosteal reaction and soft tissue mass. Lymphoma also may have low signal intensities on MRI scans similar to a fibrous tumour. Although the histologic examination of primary lymphoma of bone may show fibrous areas, it shows predominantly lymphoid tissue and only rarely areas of inflammatory infiltrate.⁷

Another differential diagnosis is eosinophilic granuloma which is comprised of clusters of mononuclear histiocytes admixed with eosinophils. Eosinophilic granulomas may have a prominent fibrous component if there has been a fracture through the lesion. However, even in this instance, the eosinophilic infiltrate is evident.¹⁵ Other uncommon differential diagnoses are haemangioma, bone cyst, chondromyxoid fibroma, non-ossifying fibroma etc.

Treatment modalities

As it is a rare lesion, treatment modalities were described by numerous surgeons as case reports or small series. Gebhardt *et al* reported on eight patients, six of whom were treated initially by intralesional curettage.¹⁷ In the eighteen patients reported by Taconis *et al*, the two local recurrences were seen in patients treated by intralesional curettage. Therefore, to decrease the risk of local recurrences, an attempt should be made to attain wide margin at the time of surgery, especially if the tumour is in an expendable bone. However if wide resection of the lesion would result in significant functional deficit, intralesional curettage may be attempted.¹¹

Raatikainen *et al* reported a case of desmoplastic fibroma in the calcaneus treated by curettage and bone grafting. No local recurrence was seen clinically or radiographically at 5 years follow-up.⁸ Nisihida *et al* reported on five patients, two with lesions in the pelvis and three with lesions in long bones, treated with curettage and bone grafting. There were no local recurrences.¹⁸ There are no series reporting the use of local adjuvant therapy in the treatment of desmoplastic fibroma. There are limited data on the effectiveness of radiation therapy. Sanfilippo *et al* reported a case of desmoplastic fibroma treated by radiation therapy with no evidence of progression for a period of 30 months.⁹

Learning points

- Desmoplastic fibroma a known metaphyseal lesion may also occur in diaphysis under rare circumstances.
- Wide excision successfully eradicates the pathology with no recurrences as noted in literature.

Competing interests None.

Patient consent Obtained.

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