Case Report and Review of The Literature

Proximal humerus chondroblastoma with a secondary aneurysmal bone cyst

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Case report

An 18-year-old man presented in our clinic with a painful shoulder that started spontaneously. He described the pain as nocturnal and not aggravated by manual labour. The pain was relieved with analgesia. On examination he had full range of movement and no local tenderness or swelling. X-rays revealed a lytic lesion in the proximal humerus *(Figure 1)*. The X-rays were reported on – a lytic lesion in the right humeral epiphysis. Differential diagnosis includes a simple bone cyst or aneurysmal bone cyst but the position was indicated as atypical. An MRI was recommended. The MRI revealed a single, well-circumscribed lesion with its epicentre in the epiphysis and stretched to the metaphysic. Bone marrow oedema was present *(Figure 3 and 4)*. The differential diagnosis was now chondroblastoma or simple bone cyst. The decision was made to first observe the growth of the lesion with serial X-rays twice a year. On the first 6 months' follow-up the lesion had grown substantially bigger, making us concerned about a pathological fracture *(Figure 2)*.

In retrospect, after the MRI report, we should have done a biopsy – especially with the number of differential diagnoses highlighted and the night pain the patient complained about.

The decision was made to surgically curettage the lesion, send the lesion for histology and fill the defect with autologous bone *(Figure 5)*. In retrospect, a frozen section should have been done. We did not have frozen section available to us in our secondary centre. We should have referred the patient to a tertiary centre for the frozen section and further management.

Intra-operatively, the lesion was very vascular but the operation was completed with no complications. Bone was taken from the iliac crest and placed in the defect. The curettaged fragments were sent for histology. The histology revealed epithelioid chondrocytes with prominent call borders and basophilic cytoplasm. The calcification of their membranes was in a chicken-wire fashion. There were also cysts filled with blood and lined by mononuclear and multinucleated cells. There was also osteoid deposited underlying the cells of the cysts. There were areas of collagenised stroma containing spindle-shaped bland cells; no necrosis was visible. The histology thus showed a chondroblastoma with a secondary aneurysmal bone cyst (solid variant). The patient did well post-operatively and maintained the full range of motion of the shoulder, good integration of the bone graft and no pathological fracture *(Figure 6)*. The shoulder is now pain free. He is currently being followed up 6-monthly to pick up any recurrence.

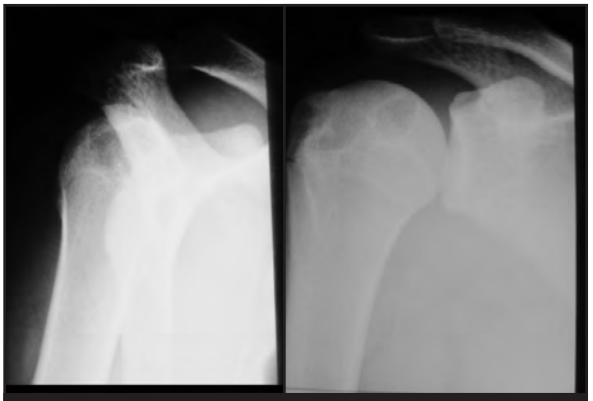

Figure 1. Initial right shoulder X-ray

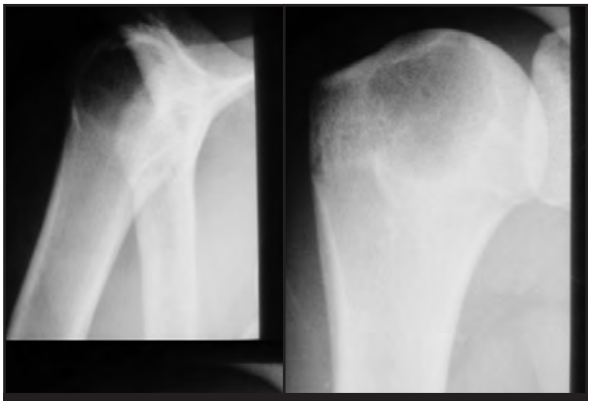

Figure 2. X-ray 6 months later

Discussion

Chondroblastomas are rare representing less than 1% of all benign bone tumours[1-16] but one author did have the incidence at 5%, possibly only including paediatric bone tumours.[17] They arise in the epiphyseal areas of long bones as well as in the apophyses.[2-5,7,9-21] Some then extend into the metaphysis.[2,3,13,21,22] They occur in the second decade – 95% of cases between 5 and 25 years of age.[2,6,8,9,11,12,15,21] They appear in males with a ratio of 2–3:1.[1,4-11,15-18]

They occur mostly in the proximal tibia,[7] proximal humerus and proximal femur[13,15] as well as in flat bones (34%).[3] Seventy-two per cent are found in the lower extremity – 50% around the knee[9,19] and 33% in the femur.[4] Twenty per cent are found in the humerus[9] – 90% of which are in the proximal humerus.[4,7] Ten per cent are also found in the small bones of the hands and feet.[19] It is the most common tumour of the patella.[19] Some say the most frequent site is the proximal humerus.[6,11] Some also describe the upper end of the femur as the most common site.[21]

Chondroblastomas often present with pain as the main symptom;[6] as well as localised tenderness, restricted painful movements, muscle atrophy, palpable mass or soft tissue swelling and joint effusion.[1-5,7-9,11-17,19,20] These symptoms are usually present for a few months.[2]

Chondroblastomas have been linked to the abnormal chromosomes 5 and 8 and p53 mutations.[1,12] Abnormalities of 8q21 predict aggressive behaviour.[12]

Chondroblastomas are associated with secondary aneurysmal bone cysts – 20%[1,7,17] to 33%.[5] They occur in abnormal bone due to haemodynamic changes.[23]

The X-ray depicts a benign, slow-growing oval or round lytic lesion eccentrically situated in part of the epiphysis. The lesion is well defined with a sclerotic margin.[2,4,7,11,16]

The authors are divided when it comes to periosteal reaction – some said it was rare[2,3,20] while others said it was common. The longer a lesion is present, the more likely periosteal reaction and matrix mineralisation is.[4] The lesion can involve other secondary ossification centres.[5] As the lesion increases in size it extends into the metaphysis. The X-ray picture is said to be diagnostic; especially involving both sides of an open growth plate.[11,12,19] The X-ray picture is divided into three groups: I – open epiphysis; II – closing plate; and III – closed plate.[17]

Bone marrow oedema seen on MRI. CT scan is used to define the cortical erosion and matrix mineralisation,[11] soft tissue extension and to evaluate aggressive and recurrent tumours.[4,9] CT scan also shows eggshell rim calcification and evaluates whether the subchondral plate of bone has been destroyed or is intact.[9]

MRI is used to evaluate transphyseal and or transcortical extension.[4] MRI also depicts any associated synovitis; bone marrow and soft tissue oedema; joint effusion; periosteal reactions and any cystic regions.[8-11]

Extensive bone marrow oedema relative to a small lesion on a MRI is seen to be benign. Presence of oedema post-operatively may be used in detecting recurrence.[24]

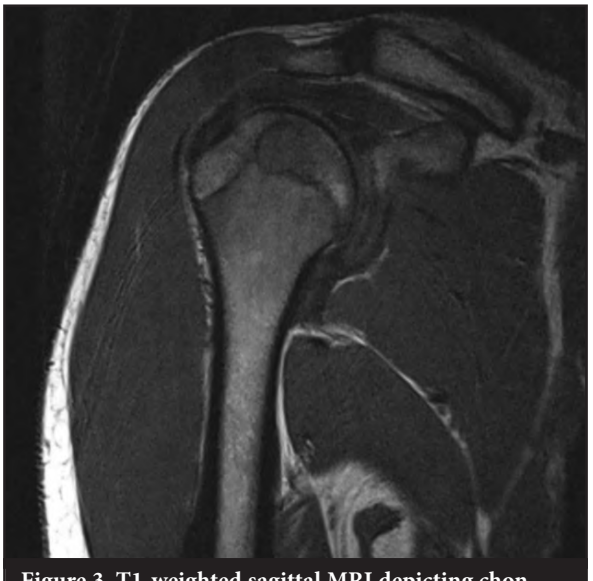

Figure 3. T1-weighted sagittal MRI depicting chondroblastoma

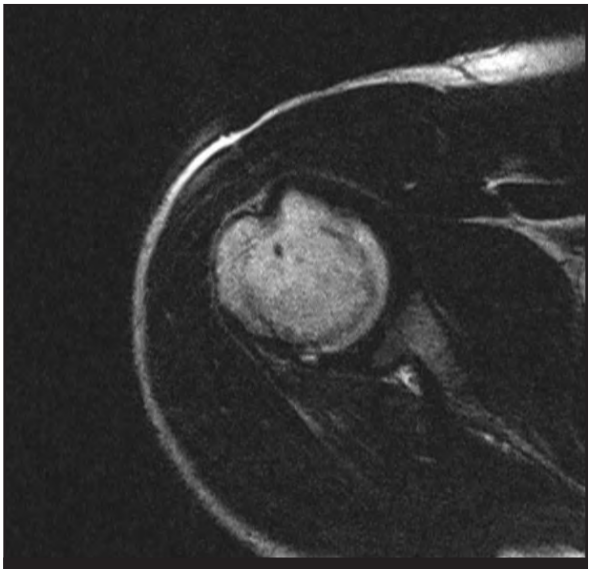

Figure 4. T1-weighted axial MRI depicting the tumour

Both can show fluid–fluid levels seen in aneurysmal bone cyst changes.[4,11]

Bone scan can show biological activity but is not very useful.[1,8]

The differential diagnosis includes chondromyxoid fibroma; chondrosarcoma; giant call tumour;[1,2,4,8,9,11,19,20,23] eosinophilic granuloma; haemangioma; osteomyelitis;[4,8] gout; rheumatoid arthritis;[8] osteoblastoma;[9,23] subchondral cyst; intraosseous ganglion;[9] enchondroma.[20]

The diagnosis is confirmed on histology by frozen section[1,11,17] – these are lobulated tumours; greyish pink soft tissue with bluish chondroid tissue with calcifications.[1,8,17,21] Haemorrhagic cystic areas are common. Secondary aneurysmal bone cysts occur in 20–25% of cases. These are then referred to as cystic chondroblastomas. Immunostaining shows S100 protein positive in mononuclear cells and stain positive for vimentin.[1,9,11] Microscopically, polyhedral cells with round-to-oval nuclei are present; honeycombed appearance and multinucleated giant cells are present.[2,6-9,11,14,17,21] Normal-appearing mitoses with calcific depostis;[3] dense eosinophilic matrix; coarse calcifications in chicken-wire pattern; cytologic atypia was rare.[4,5,7-9,11,21]

Chondroblastomas are divided into three lesion types: latent (well defined, with a complete reactive rim of bone around the lesion); active (incomplete rim) and aggressive (poorly defined margin with minimal or absent intraosseous or periosteal reaction). The aggressive lesion is active on bone scans.[17]

Chondroblastomas do not resolve spontaneously and thus treatment is needed and a few options are available.[1] Percutaneous radiofrequency ablation has been suggested. It can be used on non-weight-bearing surfaces.[1,8,9,14] The most recommended and most used mode of treatment is surgical curettage without bone grafting in older patients[4,12] or the use of polymethylmethacrylate (PMMA) in skeletally immature patients[4] or fat implantation.[1,2,5,7-9,12-15,17,19] A large window in the cortex should be made and intra-articular exposure done if needed.[6,11]

PMMA should also be used in cases of recurrences and where the tumour is likely to recur, for example in cases with secondary aneurysmal bone cysts.[4] PMMA is thought to cause heat and thus destroy residual tumour.[7] Cryotherapy with liquid nitrogen or chemical cauterisation (phenol) has also been described[1,11,14] and is said to decrease recurrence.[9] Local excision had been described.[2] Old methods of treatment included radiation, but this led to malignant transformation (chondrosarcoma, osteosarcoma or fibrosarcoma[9]) years later and is thus only recommended in areas where curettage is impossible.[2,3]

Recurrence is termed as failure of symptoms to resolve or they have returned; X-ray shows that the curetted lesion failed to fill in and consolidate or the MRI shows increased bone destruction and florid perilesional marrow oedema with or without effusion.[14]

Recurrence has an incidence of 10–45%.[1,3,5,7-9,11,12,14,15,17] It seems to be higher in flat bones and recurs on average 3 years post initial presentation so monitoring for several years is mandatory.[1,3]

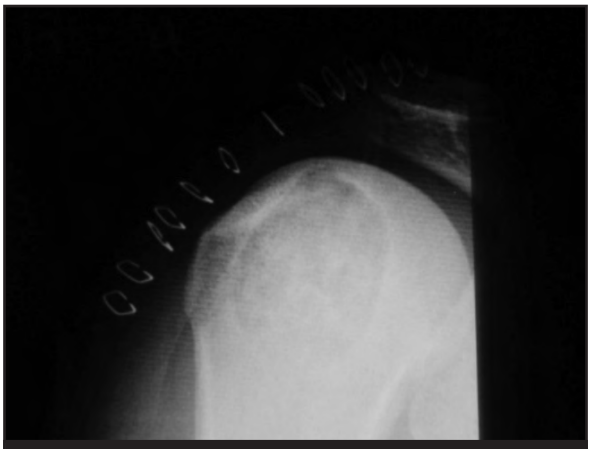

Figure 5. Post-op after curettage and bone graft

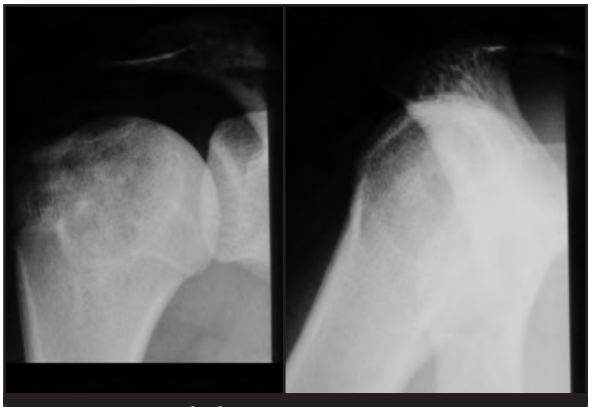

Figure 6. Six weeks later

Complications of the treatment include recurrence, post-operative infection, degenerative joint changes, fractures (although rare[4,20]), failure of the allografts, intraosseous ganglion, avascular necrosis, premature physeal closure leading to limb length discrepancy and malignant transformation.[1,4] The risk of recurrence has been linked to several factors but authors are still divided on some: some say they are not related[15] to age (younger do better says some authors[13,14]); sex; size of the tumour (some authors said it was[4,7,10]); amount of calcification or vascular invasion or duration of follow-up or method of treatment (bone grafting vs phenol showed no difference[3] but inadequate surgery showed increased recurrence[13]) but were related to open physeal plates; because the curettage is often less aggressive to try and preserve the growth plate.[1,5,7]

One author feels the growth plate is actually resilient and aggressive curettage is appropriate and they get good functional results.[13,14]

One author found the recurrence 100% when the tumour is associated with aneurysmal bone cyst[3-5,13] but some authors reported that this is not confirmed.[1,3] Some authors found an increased risk of recurrence in tumours in the proximal femur and greater trochanter, possibly due to the difficulty of gaining access.[4,5,7,13,19] Prognosis is excellent, with nearly normal to normal function.[6,19]

Treatment of recurrences is repeat curettage.[1] Patients with local recurrence should be screened for metastases yearly by way of a chest X-ray.[14]

There is an aggressive type of chondroblastoma with benign histology but the tumour is of an increased size and it metastasises to the lungs and the surrounding soft tissue. These metastases can occur up to 30+ years after the initial tumour.[1,4,8,10,12,17] This type is likely to recur.[9,13]

Malignant transformation can also occur up to 10 years after the initial benign lesion and can occur with pulmonary metastases. The histology is similarly benign but has abundant and abnormal mitotic figures, tumour necrosis and intravascular thrombi. These tumours have a poor prognosis.[1,8,13]

Conclusion

One should always have a high index of suspicion of chondroblastoma when a young healthy male with long duration symptoms has pain and a tumour in the epiphysis of a long bone.[2] Early intervention – when there are multiple differential diagnoses and night pain as well as frozen section before definitive treatment – is good clinical practice.

No benefits of any form have been received from a commercial party related directly or indirectly to the subject of this article.

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