Extraosseous Giant Cell Tumour of the pinna presenting in a child: Case report and review of the literature

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Citation

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Abstract

Background: The literature on extraosseous giant cell tumours (GCT) of the soft tissues is relatively sparse.Methods: We present a previously unreported case of GCT of soft parts with two unique characteristics, which distinguish it from previous reports in the literature.Results: The first: this case involves the pinna of a child, GCT has been previously documented in the head and neck, but not involving the ear, especially in a young child. The second: the benign nature of the disease despite the presence of an abnormal mitotic figure which is generally associated with the malignant giant cell tumour variant and a more aggressive disease.Conclusion: We discuss this rare condition and the literature is also reviewed.

INTRODUCTION

Giant cell tumours (GCT) of bone are benign, locally invasive lesions with a high rate of recurrence, accounting for approximately 5% of all primary bone tumours. [1] They typically occur in the metaphysis and epiphysis of long bones as an osteolytic tumour in a skeletally mature individual. [2] Two per cent arise in the head and neck area. [3] Although the recurrence rate following excision or curettage is high, [4, 5] only 1.5% of the cases actually recur within the adjacent soft tissues. [6]

Primary giant cell tumours that are histologically indistinguishable from giant cell tumours of bone may also rarely arise in soft tissue. [7] This tumour is classically composed of a monotonous proliferation of mononuclear cells and osteoclast-like giant cells immersed in a richly vascularised stroma that are homogenously distributed throughout the tumour. [8, 9] Metaplastic bone trabeculae are also a feature of these tumours. [7]

There has been controversy in the literature regarding the nomenclature and the existence of a benign and a malignant form of the condition, based on the clinico-pathological features. This is illustrated by the variety of different names in the literature including ‘extraosseous giant cell tumour of soft tissue’, ‘primary giant cell tumour of soft tissues/parts’, ‘giant cell tumour of low malignant potential’ and ‘malignant giant cell tumour’.

They are most prevalent in the superficial or deep soft tissues of the extremities. Other reported sites include the paravertebral soft tissues [10], parotid and other major salivary glands [11] and parenchymal organs such as the pancreas, thyroid, lung, breast, ovary and kidney. [11, 12] A case of a giant-cell extraosseous tumour of soft tissues has even been reported in the peritoneal cavity of a four year old girl. [13]

We present the first reported case of an extraosseous giant cell tumour of the pinna in a child, with distinguishing histological features.

CASE REPORT

A 10 year-old female presented to the otolaryngology outpatient department with a three-week history of an almond sized lump of the left pinna. She was otherwise well in herself and had no other history of note. On examination she was found to have a 1.5 x 1.5 cm lump on the posterior surface of the left pinna. She was listed for surgical excision of the mass.

Three months following her first appointment she was seen in the preadmission clinic and a dramatic increase in the size of the lump had occurred: the lump having doubled in size. It also appeared to be more solid and vascular on closer inspection.

At Operation, a solid mass was excised, this was adherent to the pinna superiorly and after excision, the area of cartilage

involved was curetted. After an uneventful recovery she was discharged home the following day.

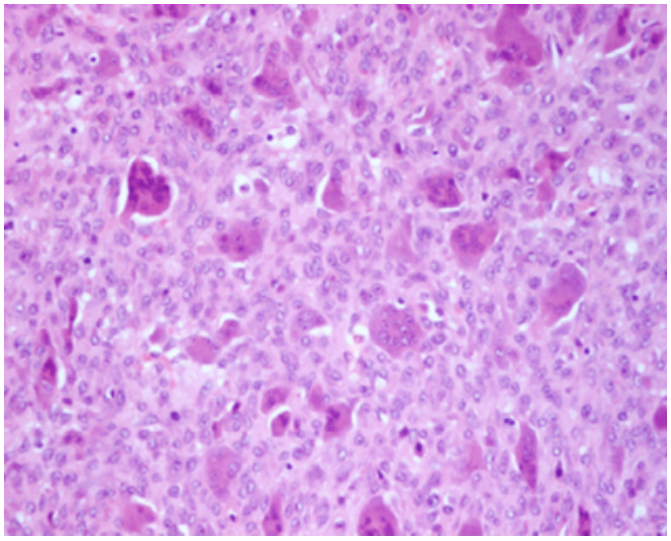
HISTOPATHOLOGY

Macroscopically the tumour was composed of firm tan tissue; two specimens of 15x10x6 mm and 35x20x10mm were obtained.

Microscopy showed a non-capsulated hypercellular tumour extending to the margins of excision. The tumour showed a uniform appearance in all sections examined and consisted of an almost evenly distributed pattern of osteoclast-like giant cells in a background of rather monomorphic mononuclear cells (Figure 1).

Figure 1

Figure 1. The tumour demonstrating a uniform appearance in all sections examined and consisting of an almost evenly distributed pattern of osteoclast-like giant cells in a background of rather monomorphic mononuclear cells (Figure 1). Magnification x

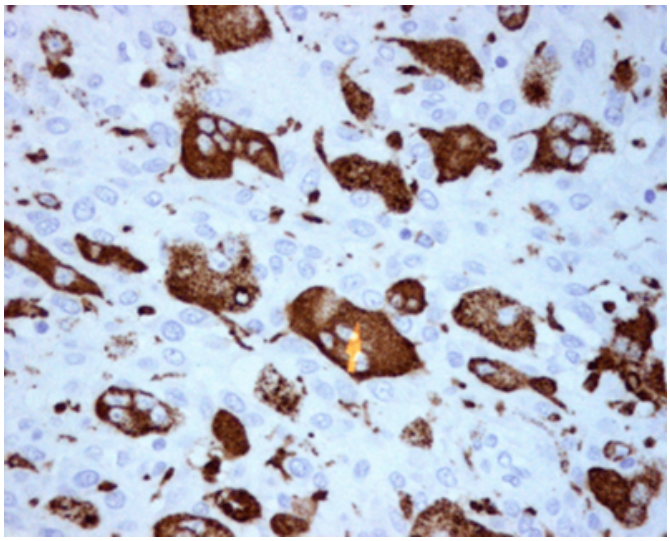


The mononuclear cells had vesicular nuclei and nucleoli and indistinct cytoplasmic margins. Focally the background matrix appeared to be less cellular and hyalinised but no osteoid or cartilaginous components were seen. The tumour was mitotically active and an abnormal mitosis was identified. Intravascular invasion by the tumour cells was also noted.

Immunohistochemistry showed positivity of the mononuclear cells for desmin, alpha smooth muscle actin and smooth muscle myosin but this was considered as non-specific staining. The osteoclast-like giant cells also stained positively for these markers but also showed positivity for a macrophage marker CD68 (Figure 2). The tumour stained negatively for CAM 5.2, CD34, CD31 and Factor 13A.

Figure 2

Figure 2. Osteoclast–like giant cells staining positively for markers also showing positivity for macrophage marker CD68 (Figure 2). Magnification x20



The morphological appearances of the tumour were consistent with a rare soft tissue tumour known as an extraosseous giant cell tumour. Due to the rarity of this tumour in this age group and its unusual location slides were sent for expert opinion. The outcome was that this was indeed an extraosseous giant cell tumour. Five years post excision, there has been no evidence of recurrence.

DISCUSSION

Giant cell tumours of soft parts were first distinguished from other giant cell containing tumours by Salm and Sissons in 1972, in their 10 patient series. [14] This was followed by Guccion and Enzinger's larger study of 32 cases which recognised the malignant potential of these tumours. [15] This division was included in the World Health Organisation (WHO) Classification of Tumours of Soft Tissue and Bone in 2002. [16] The more benign form, with absence or little cytologic atypia and low to moderate mitotic activity without atypical figures, are considered giant cell tumours of soft tissue/ soft parts or low malignant potential whereas the deeper, histologically high-grade tumours are documented as malignant giant cell tumours of soft parts. [17, 18] The latter, more aggressive, entity having metastatic potential. Folpe concluded however, that to label some as benign and some as malignant oversimplifies the problem as even those of low malignant potential may produce metastases. [17, 19]

Grading the condition also produces controversy as illustrated in giant cell tumour of bone. [20]

PRESENTATION

Giant cell tumour of soft parts usually presents as a mass developing over a short time frame (less than one year), although it may progress over 15 years. [14] Tumours tend to be multinodular and can occur in superficial or deep locations. Deeper tumours are more likely to be malignant giant cell tumours. [17, 21] There may be a preceding history of trauma leading to the lesion being misdiagnosed as a haematoma. [15] Past reviews have documented that only 5 of the reported 78 tumours were in the head and neck, the majority (70%) being in the extremities, with the thigh being the most common site. [9, 14, 15, 17, 22,] Previously documented head and neck sites include the forehead [18] and nasal cavity. [23]

The age range of patients with primary GCT of soft tissue is 1-86 years with a female preponderance of 3/2. [9, 14, 17] Most cases occur in middle-aged and older patients, although rarely it has been described in children. In a study of 22 cases only six patients were under the age of 20 years. [9] The mean age of patients in Guccion's study, which consisted of more malignant tumours, was 56. [15] The size of lesion has ranged from 1 to 10cm diameter in the larger studies, although Galed-Placed reported a tumour measuring 20x8cm. [24]

MACROSCOPIC/ MICROSCOPIC FINDINGS

Macroscopically these tumours tend to be fleshy, rubbery, red-brown or grey on their cut surfaces. [9] The histopathological findings in this case were similar to those reported in the literature. A monotonous proliferation of mononuclear cells and osteoclast-like giant cells immersed in a richly vascularised stroma that are homogenously distributed throughout the tumour is a common finding in these tumours. [8, 9] These neoplasms are frequently multinodular and mitotic activity is typical in the morphology. [9] As was reported in our case, vascular invasion was identified in seven (31.8%) of patients in a study of 22 cases. [9]

Giant cell tumours of soft tissue show nuclear mitosis but their atypia is mild to moderate. Pleomorphic giant cells are absent. In contrast, malignant giant cell tumours are identified by their characteristically malignant features: nuclear enlargement, nuclear irregularity and pleomorphism, prominent nucleoli, necrosis and atypical mitotic figures. [9, 17, 23] Our case identified an abnormal mitotic figure, however the nuclei were vesicular.

Immunohistochemical findings that correlate well with the diagnosis of GCT of soft tissue include CD 68 expression as occurred in the current case. [9] However in previous case series the tumours have lacked staining for desmin, in contrast to our case, as well as CD31 as was the case in our patient. [9] CD68 and SMA staining are not common in malignant giant cell tumours. [25]

Cytogenetic analysis of all excised tumours has been recommended. [26] These suggestions being based upon the prior documentation that 75% of osseous GCTs share various cytogenetic abnormalities, in which telomeric association was the most common abnormal chromosomal finding. [26] Indeed, it was present in nearly all recurrent and metastatic tumours. In contrast, only approximately 25% of GCTs with indolent behaviour demonstrated this chromosomal abnormality. [26] Such studies involving GCT of soft parts are lacking.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of primary GCT of the soft tissues at the histologic level includes a variety of benign and malignant entities rich in multinucleated or osteoclast-like giant cells. [27] First, distinction from soft tissue extension of its bony counterpart must be ensured clinically or radiographically. In the case described this was easily achieved due to its location in the pinna. [27] Giant cell malignant fibrous histiocytoma, osteoclast-like giant cell rich leiomyosarcoma, and extraskeletal osteosarcoma are usually largely deeply seated lesions with obvious cellular atypia. [27] In contrast primary GCT of the soft tissues is frequently superficial and devoid of significant pleomorphism and atypical mitoses. The more malignant form of Giant cell tumour is differentiated from malignant fibrous histiocytoma by its vascularised stroma composed of round or oval mononuclear cells, the uniform distribution of the osteoclast-like giant cells and lack of xanthomatous cells. [9, 15]

Epitheliod sarcoma may be considered, however epitheloid sarcomas show larger tumour cells, zones of central necrosis, and a more infiltrative pattern. [27] Other histologically similar tumours include atypical fibroxanthoma, plexiform fibrohistiocytic tumour and benign tumours such as cellular dermatofibromas. [27] Giant cells may be an inconsistent feature in giant cell rich leiomyosarcoma, epitheliod sarcoma

and malignant mesenchymoma as opposed to a definitional feature in this condition. [17] Giant cell tumour of soft parts has no relationship to synovial tissue, and lacks the varied cell population of giant cell tumours of tendon sheath, which co-express CD68 and CD45. [17]

PROGNOSIS

The prognosis of patients with primary GCT of the soft tissues was originally thought to be intermediate and vary with depth. Guccion and Enzinger proposed that deep tumours were more likely to have an aggressive clinical course with the development of metastases being more common than those with superficial tumours. [15] Here, 15 of 20 patients with deep tumours developed pulmonary metastases, even in cases where amputation had been performed. Oliveira's review of 22 cases of GCT of soft tissues reported one of 13 patients (8%) who were followed up for periods ranging from 2 months to 130 months with superficial tumours developed metastases and died of tumour 12 months after initial treatment. [9, 28] The death rate for the 3 deeply located GCT of soft tissue was 0%. [9, 28] In contrast, all 10 patients in the original GCT series by Salm and Sissons, over an average 4-year follow-up period, had a benign clinical course with no metastases. [14]

Following the separation of primary GCT of soft tissue from malignant giant cell tumour of soft parts, it has been shown that the depth of lesion is a less significant diagnostic factor than histological features and that the majority of primary GCT of soft tissues have a benign clinical course. [9, 17] Neither lymphovascular invasion, nor a high mitotic rate, appear to affect prognosis however. [9, 17] The more malignant tumours tend to require more extensive resections, even amputations, have a higher recurrence rate and have a higher propensity for metastases. [9, 15, 27] The best therapeutic approach for those of low malignant potential is surgical resection with margins free of tumour, although the recurrence rate appears to be approximately 15%. [18] In this case, one margin was positive but the cartilage of the pinna was carefully and thoroughly debrided with a curette after careful removal of the tumour to obtain good surgical clearance with a good cosmetic result. Hence we believe the actual area was free of residual tumour. As is recommended in the literature this young girl will be followed up and monitored for signs of recurrence and/or metastasis.

CONCLUSION

GCT of the pinna of in a child, of benign nature despite the presence of an abnormal mitotic figure which is generally associated with the malignant giant cell tumour variant and a more aggressive disease, has not been reported in the literature before. We discuss this unique case and this condition.

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