Primary Liposarcoma of Temporal Bone in a Child

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Abstract
Liposarcoma is a malignant tumour of soft tissue and the temporal bone is an unusual location for this tumour, especially for children. A boy suffered primary liposarcoma of temporal bone causing headache and vomiting. Noncontrast CT and MRI scan of the head showed well defined mass in the left temporal bone. Gross total resection was performed. Histopathological examination revealed a myxoid liposarcoma. The postoperative course was uneventful. The patient is now disease-free at 4.5 years follow-up. The presentations, imaging findings, and progress were reported.

Key words Liposarcoma; Primary; Temporal bone

Introduction
Primary liposarcoma of temporal bone is uncommon in children. Surgical excision is the main form of therapy. A boy presenting headache and vomiting was diagnosed to have a primary liposarcoma of temporal bone. The lesion was gross totally resected and the patient enjoys along disease free remission.

Case Report
A 10-year-old boy presented with 15 days history of headache and vomiting which more severe at night. His physical examination was unremarkable except for a left temporal region painless swelling. It was non-tender with normal overlying skin. He denied having any local trauma and there were no neurological deficits. Routine blood examination was essentially normal. Noncontrast CT scan of the head with bone window showed 3 cm x 4 cm x 5 cm, well defined mass of low attenuation situated in the left temporal bone. The mass had punctuate calcifications (Figures 1A & 1B). Noncontrast MRI scan of the head showed 3 cm x 4 cm x 5 cm, well defined mass of few high-signal intensity foci within low-signal intensity mass on T₁-weighted images, and high-signal intensity mass on T₂-weighted images in the left temporal bone (Figures 1C & 1D). With the clinical impression of left temporal bone neoplasm, gross total resection was performed. Histopathological examination revealed a myxoid liposarcoma (H&E, x 200) (Figure 2). The postoperative course was uneventful. The patient is now disease-free for 4.5 years after the surgery.

Discussion
Liposarcoma is a malignancy of adipose cells. In adults, it is one of the most common soft-tissue sarcomas, accounting for 9.8-16% of all soft-tissue sarcomas with an annual incidence rate of 2.5/million. It’s commonly found in the retroperitoneal cavity or extremities (particularly the...
thigh). Only 3% of liposarcomas occur in the head and neck region (usually at the neck or cheek). Liposarcomas are slightly more common in males but has no ethnic predisposition. It was most commonly found in the fifth to seventh decades, but can occur in children occasionally. The youngest patient reported that we could found in MEDLINE was a 3-year-old boy with an intrathoracic tumour.

Liposarcoma is a lipogenic tumour of large deep-seated connective tissue spaces. It is composed of different biological subtypes, including low and high grades. On the basis of histological appearances, World Health Organization classification divided liposarcomas into 5 categories of, namely: (1) well differentiated, including the adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell and (5) pleomorphic types. Myxoid liposarcoma is the commonest type of liposarcoma, accounting for 50% of all cases. Cytogenetic studies of myxoid liposarcoma revealed a highly specific gene translocation, t(12;16)(q13;p11). It results in a fusion transcript between of FUS gene on chromosome 16 with the CHOP gene on chromosome 12. Both genes probably play a significant role in adipogenic differentiation. Other complex and rare translocation variants involving either 12q13 or 16p11 have also been reported.

On imaging, most liposarcomas have well-defined and lobulated margins, composed of mainly fat with septa or nodules. The appearance of liposarcomas on MRI reflects its degree of differentiation. Many of these tumours contain fat and it can be characterised with MRI because of their similar signal intensity to that of subcutaneous fat. As in our case, the few high T1-weighted signal intensity foci within low-signal intensity mass usually represents fatty. Whereas the high T2-weighted signal probably represents the predominant myxoid matrix. Although the MRI appearances of this tumour are quite typical, it should be differentiate from other soft tissue tumour such as lipoma, neurofibroma, angiofibroma, rhabdomyosarcoma, and fibrous histiocytoma.

In terms of treatment, surgical excision remains the main form of therapy. Local radiation and/or chemotherapy may be helpful for high-grade tumours. In general, the prognosis is related to the histological types. For example, well-differentiated and myxoid types have a 100% and 88% 5-year disease free survival rates respectively. However, tumours with poorly circumscribed margin or those recur locally after excision dose not do well, even they rarely metastasize. Repeated local recurring tumours may finally evolve into a higher grade of sarcoma with metastasis potential. The misdiagnosis of a liposarcoma as other benign tumour therefore should be avoided.
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References