Sclerotic Liposarcoma in the Deep Temporal Space of a 7-year-old Child

Wei-Li Kong, Wen Li

Department of Otolaryngology, Head and Neck Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan 610041, China

To the Editor: Liposarcoma is the most common soft-tissue sarcoma and accounts for 20% of all mesenchymal tumors. It usually occurs in the deep soft tissue of extremities and retroperitoneum in adults.[1] Apart from the aforesaid locations, the sarcoma also occurs in sites such as orbit, neck, oral, paratesticular, and salivary gland regions. The number of the cases under the age of 10 years that can actually be traced has not yet exceeded five in literature.[2,3] Herein, we report an extremely rare case of sclerotic liposarcoma that has occurred in the deep temporal space in a 7-year-old patient.

A 7-year-old girl, with an intumescent temporal region, was admitted to the Department of Otolaryngology, Head and Neck Surgery, West China Hospital in December 2016. From her medical history, we learned the information that a painless horsebean-shaped mass had emerged in the right temporal region for 2 months and had been progressively expanding in size during the last month. T1-weighted magnetic resonance imaging demonstrated slightly increased signals with the dimensions 3 cm × 3 cm in the right temporal region; no obvious bony destruction was detected. Surgical exploration revealed a smooth, capsulated mass, tightly jammed between the squamous bone and the temporal muscle. The tumor was completely excised. Gross anatomy observation revealed that the surface and cross section of the tumor strikingly resembled a typical mixed tumor of salivary gland. Postoperative histopathological study revealed scattered bizarre stromal cells and lipoblasts with lipid droplets within the cytoplasm displacing the nucleus to the side on the hyperplastic fibrous tissue background. Immunohistochemically, the tumor cells were positive for vimentin, sex determining region of Y chromosome-related high-mobility group box gene-9 (SOX-9), S-100, P16, murine double minute 2 (MDM2), and cyclin-dependent kinase 4 (CDK4) and negative for pan-cytokeratin (PCK), epithelial membrane antigen (EMA), smooth muscle actin (SMA), desmin, myogenin, cluster of differentiation 10 (CD10), CD31, CD34, caldesmon, CD99, beta-catenin, signal transducer, and activator of transcription 6 (STAT-6). Molecular immunology Borstel number 1 (MIB-1) antibody-positive cells accounted for 1% (MXB Biotechnologies, Fuzhou, China). *MDM2* gene (12q13-15) amplification was proved by fluorescence *in situ* hybridization. The diagnosis of sclerotic liposarcoma was finally established [Figure 1]. The patient was eventless postoperatively.

Access this article online	
Quick Response Code:	Website: www.cmj.org
	DOI: 10.4103/0366-6999.209907

Liposarcoma is rare in children and almost never found in patients younger than 8 years old. Previously reported liposarcoma cases in infancy are mostly deemed as lipoblastomatosis now.[2,3] Normally, liposarcoma rarely occurs in head and neck and unlikely causes

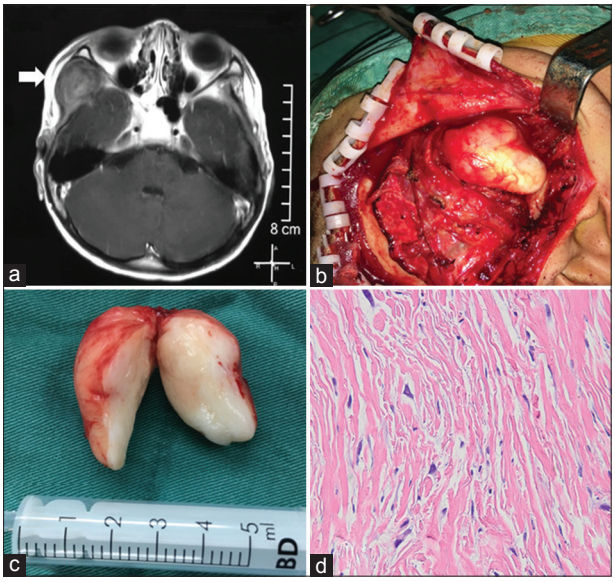

Figure 1: MRI, operative view, and histopathology of the sclerotic liposarcoma. (a) T1-weighted MRI revealed slightly increased signals in the right temporal region without obvious bony destruction (white arrow). (b) Operative field demonstrated capsulated tumor. (c) The surface and cross section of the tumor resembled a typical mixed tumor of salivary gland. (d) Histopathological examination demonstrated scattered bizarre stromal cells and lipoblasts with lipid droplets within the cytoplasm displacing the nucleus to the side of the cell (hematoxylin-eosin staining, original magnification, ×400). MRI: Magnetic resonance imaging.

Address for correspondence: Prof. Wen Li, Department of Otolaryngology, Head and Neck Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan 610041, China
E-Mail: church.ent.wc@163.com

Received: 07-04-2017 **Edited by:** Peng Lyu
How to cite this article: Kong WL, Li W. Sclerotic Liposarcoma in the Deep Temporal Space of a 7-year-old Child. Chin Med J 2017;130:1759-60.

obvious manifestations at early stage. Only when the tumor expands up to a certain size, the symptoms such as pain or functional disturbances occur. Retroperitoneal tumors might emerge with signs of weight loss and abdominal pain. Histologically, liposarcoma can be classified into five types, namely, mixed, myxoid, pleomorphic, dedifferentiated, and atypical lipomatous types. In this case, when abundant fibrous collagen accumulates in the tumor, multifocal sampling is required in case that limited lipomatous elements are neglected.

Currently, no consensus has been reached on the prognosis of liposarcoma yet. It might correlate to the pathological subtype, location, and extent of resection. It seems that as for liposarcoma in head and neck, disease-specific survival and overall survival are significantly greater than that of the sarcomas which occur in other regions.[1,4] Surgical complete excision is the mainstay and is rational for the prevention of recurrence and dedifferentiation, though dedifferentiation will not necessarily occur synchronously with recurrence. The effectiveness of radiotherapy and chemotherapy remains a widely controversial topic due to the paucity of documented cases.[5]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's guardians have given their consent for the patient's images and other clinical information to be reported in the journal. The patient's guardians understand that names and initials of the patient will not be published and due efforts will be made to conceal the identity of the patient, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Gerry D, Fox NF, Spruill LS, Lentsch EJ. Liposarcoma of the head and neck: Analysis of 318 cases with comparison to non-head and neck sites. Head Neck 2014;36:393-400. doi: 10.1002/hed.23311.
2. Alaggio R, Coffin CM, Weiss SW, Bridge JA, Issakov J, Oliveira AM, *et al.* Liposarcomas in young patients: A study of 82 cases occurring in patients younger than 22 years of age. Am J Surg Pathol 2009;33:645-58. doi: 10.1097/PAS.0b013e3181963c9c.
3. Okamoto S, Machinami R, Tanizawa T, Matsumoto S, Lee GH, Ishikawa Y. Dedifferentiated liposarcoma with rhabdomyoblastic differentiation in an 8-year-old girl. Pathol Res Pract 2010;206:191-6. doi: 10.1016/j.prp.2009.05.005.
4. Fanburg-Smith JC, Furlong MA, Childers EL. Liposarcoma of the oral and salivary gland region: A clinicopathologic study of 18 cases with emphasis on specific sites, morphologic subtypes, and clinical outcome. Mod Pathol 2002;15:1020-31. doi: 10.1097/01.MP.0000027625.79334.F5.
5. Kaplan R, Bratcher GO, Freeman D, Seid AB, Cotton R. Liposarcoma of the neck. Report of a case. Laryngoscope 1981;91:1375-8. doi: 10.1288/00005537-198108000-00020.