Case Report
Plexiform fibrohistiocytic tumor in a rare location: zygomatic arch

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Abstract: A case of plexiform fibrohistiocytic tumor (PFHT) developing in a 3-year-old girl who presented with the right cheek swollen was reported. Histologically, the tumor displayed multiple nodules of plexiform distribution within the deep dermis and subdermal adipose tissue, consists of mononuclear histiocyte-like cells and multinucleated osteoclast-like cells, the nodules is surrounded by spindle fibroblast-like cells. They were positive for vimentin, SMA, MSA, and CD68, but negative for AE1/AE3, desmin, S100, CD34. This case demonstrates that PFHT is a rare soft tissue neoplasm, in head and neck is more rare, various misleading morphological features should been taken into consideration in the diagnosis.

Keywords: Plexiform fibrohistiocytic tumor, zygomatic arch, soft tissue neoplasm

Introduction

Plexiform fibrohistiocytic tumor (PFHT) is an uncommon Mesenchymal neoplasm of low malignant potential, first described by Enzinger and Zhang in 1988 [1]. It usually affects children and young adolescents [2], the median age of the largest series reported was 14.5 years [3], most of them were female. The tumor is usually localized in the upper extremities, head and neck rare, and presents multinodular lesions in deep dermis and subcutaneous tissue. Here, we added a case of PFHT with uncommon location to discuss the clinicopathologic, immunohistochemical, and differential diagnosis.

Clinical history

A 3-year-old girl was admitted for the right cheek bone mass with a month, consciously increased, without pain and heat. A computed tomography scan showed a soft tissue mass of 18.5 × 16.9 mm on the right side of the front of masseter with low density, compression of adjacent cheekbones (Figure 1F). Complete resection of the mass and part of zygomatic, and postoperative patients in stable condition, without recurrence and metastasis.

Materials and methods

The surgical specimen were fixed in 4% buffered formalin, embedded routinely in paraffin and then stained with hematoxylin and eosin. Immunohistochemical use ready-to-use antibodies. The antibodies included CD68, SMA, Vimentin, S-100, CD34, Bcl-2, HMB-45, Desmin, EMA, AE1/AE3, ERG, Caldesmon, β-catenin, Ki-67. Bought used antibody and immunohistochemical staining kits from Roche co., LTD, Set up a control, regular.

Results

Grossly, the tumor measuring 2 × 2 × 1 cm, with zygomatic adhesion, revealed a firm and white-grey cut surface without distinct demarcation. Histologically, the PFHT showed a multiple nodules of plexiform distribution or consist of spindle cell bundle, sometimes small nodules can be fused (Figure 1A). The nodules are composed of plexiform proliferation of mononuclear histiocyte-like cells, multinucleated osteoclast-like cells, and spindle fibroblast-like cells components in variable proportions (Figure 1B). The mitotic activity was low (<1/HPF) and no necrosis was seen. Local area can be seen vascular (Figure 1C) tumor invasion and bone tissue invasion (Figure 1D).
The tumor cells were positive for CD68 (Figure 1E), SMA, and Vimentin, whereas negative for AE1/AE3, S-100, CD34, Bcl-2, HMB-45, Desmin, EMA, ERG, Caldesmon, β-catenin. Proliferative index Ki-67 was approximately 10%.

**Discussion**

Firstly described by Enzinger and Zhang [1]. PFHT is rare, and has been described in a smaller percentage of cases [4-6]. PFHT have been considered to have a low-grade malignant
PFHT in zygomatic arch

Immunohistochemically, the vast majority of the tumor cells show a consistent immunophenotype, including the histiocytic cells positive for CD68, and the fibroblastic cells stain for SMA, and Vimentin [3]; but absence of AE1/AE3, S-100, CD34, Bcl-2, HMB-45, Desmin, EMA, ERG, Caldesmon, β-catenin. The molecular diagnosis of PFHT was not reliable, because of lack of specific chromosomal hallmarks [12]. Several entities with the plexiform structure, giant cell structure, eomyfibrous tissue cell tumor and spindle cells tumor should be taken into consideration in the differential diagnosis. one differential diagnosis of PFHT is cellular neurothekeoma (CNT), both PFT and CNT are likely to occur in young people, and both have similar histological and immunophenotypical, some reports found that PFHT and CNT might have a common histogenesis [13]. But CNT manifest a plexiform pattern consist of nests and fascicles of eosinophilic epithelioid, multinucleated giant cells can occasionally be seen, CNT tend to have more atypical cells and a higher mitotic activity; PFHT displayed multiple nodules consists of mononuclear histiocyte-like cells, multinucleated osteoclast-like cells, and spindle fibroblast-like cells, without epithelioid cells. Other tumors with plexiform structure are easily confused with PFHT such as plexiform schwannoma, plexiform neurofibroma; tumors with giant cell structure should be as a differential diagnosis: giant cell angiolipoma, benign and malignant soft tissue giant cell tumor; some spindle-cell tumors should be identification with PFHT, such as fibromatosis, fibrous hamartoma of infancy, nodular fasciitis, dermatofibroma, deep benign fibrous histiocytoma and so on.

Effective differential diagnosis is required. PFHT is intermediate tumor, and recurrence is high without complete resection, so that some benign tumors were identified with, in this way can as much as possible reduce tumor recurrence.

In conclusion, PFHT is a distinctive rare fibrohistiocytic tumor, occurred in head and neck is rarer. There is no standard systemic treatment, at present extensive surgical resection is the main treatment, long-term follow-up is necessary to perceive any local recurrence, as well as distance metastases. To be familiar with this entity could contribute to avoid misdiagnosis when we face the similar diseases.
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Disclosure of conflict of interest

None.

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