Plexiform fibrohistiocytic tumor in the submandibular region: A rare diagnosis

Ruma Pahwa · Nita Khurana

Abstract Plexiform fibrohistiocytic tumor (PFHT) is a rare mesenchymal neoplasm of intermediate malignancy, first reported by Enzinger and Zhang in 1988 [1]. It mainly affects children and young adults and preferentially involves the upper extremity [1, 2].

We report a rare case in the submandibular region which was diagnosed on histopathology.

Keywords Plexiform fibrohistiocytic tumor · Submandibular region · Mesenchymal neoplasm

Introduction

Plexiform fibrohistiocytic tumor (PFHT) is an uncommon mesenchymal neoplasm of intermediate malignancy [2] that can cause difficulty in diagnosis and surgical management [3]. It has a predilection for children and young adults and usually involves the upper limbs as a slow growing painless mass. The tumor has a high local recurrence rate though it metastasizes only rarely [2]. We present this case for its unusual location and discuss the differential diagnosis of lesions in the submandibular region.

Case report

A young male patient presented with a painless, slow growing swelling in the submandibular region first noticed 4 months back. It was clinically diagnosed as a submandibular lymphnode. The swelling measured 2.0×2.0 cm, was present on the left side, well defined and firm. No other significant findings were recorded on clinical examination. It was enucleated and sent for histopathological examination. No surrounding tissue was included.

Grossly the tumor measured $2.0 \times 2.0 \times 1.0$ cm, was well circumscribed, grey white to grey brown, solid.

Histological examination showed a tumor comprising of spindle cells in a fascicular arrangement with a few scattered multinucleated giant cells interspersed in between. The spindle cells were seen infiltrating the subcutaneous adipose tissue. These cells showed mild focal pleomorphism and low mitosis. On immunohistochemical examination vimentin positivity and focal positivity of giant cells with CD 68 was observed.

Discussion

PFHT is an uncommon mesenchymal tumor in children and young adults [2], more frequent in females and mostly

R. Pahwa¹ · N. Khurana²

¹Department of Pathology,
Dr Baba Saheb Ambedkar Hospital, Delhi, India

²Department of Pathology,
Maulana Azad Medical College, New Delhi, India

R. Pahwa (✉)

E-mail: rumaarora1@rediffmail.com

occurs in the upper extremity followed by lower extremity and head and neck region [4, 5]. Since it was first described in 1988 [1] there have been few series [4–6] and many single case reports in literature. We present a rare case in a male patient presenting in the submandibular region.

Three main histological patterns of PFHT have been recognized- fibroblastic, histiocytic and multinucleate giant cell type. It is a poorly demarcated dermal or subcutaneous mass with multinodular plexiform growth [2, 5] and infiltrative growth pattern that can show microfat cells due to subcutis infiltration [5]. The tumor uniformly displays immunoreactivity for vimentin, focally for SMA and CD 68 and has features of myofibroblasts and histiocyte-like cells ultrastructurally [2, 5]. Our case was wellcircumscribed, predominantly fibroblastic however, showed infiltration of subcutis with presence of microfat cells within the tumor (Fig. 1). It showed immunoreactivity with vimentin and focally with CD 68.

Fig. 1 Sections shows fascicles of spindle cells infiltrating subcutaneous adipose tissue (H & E, $\times 100$). Inset shows high power view (H & E, $\times 400$) showing mild focal nuclear pleomorphism and scattered multinucleate giant cells

Differential diagnosis of PFHT includes plexiform neurofibroma, plexiform schwannoma, cellular neurothekeoma, fibrous hamartoma of infancy, deep benign fibrous

histiocytoma, dermatofibroma, benign and malignant soft tissue giant cell tumor and myofibromatosis [2]. Fibrous hamartoma and soft tissue giant cell tumor occur in adults and show solid growth pattern while dermatofibroma shows a storiform pattern. A negative S-100 helps to differentiate PFHT from plexiform neurofibroma, plexiform schwannoma and cellular neurothekeoma [2].

Surgical resection is the treatment of choice [2] however local recurrence has been reported to be between 12.5–40% [1, 5, 6]. Regional lymphnode metastases has been reported as 6% in two of the series [1, 5]. A single case of PFHT with systemic metastasis and death has been reported [7]. Hence, wide excision with long term follow-up is recommended to detect any local recurrence as well as any nodal or systemic metastasis [2].

We present this case for its unusual presentation in the submandibular region where it must be considered as a possible differential diagnosis.

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