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Plexiform Fibrohistiocytic Tumor of the Neck
- A Case Report -

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Plexiform fibrohistiocytic tumor (PFT) is a rare, low-grade soft tissue tumor that occurrs primarily in children and young adults. The most common location of PFT is the upper extremity, and there are very few reports of PFT in the neck. We report here on a case of PFT presenting as a painless subcutaneous nodule in the neck of a 46-year-old woman. Histologically, this subcutaneous tumor was composed of a plexiform proliferation of histiocyte-like cells and fibroblast-like cells along with a few multinucleated osteoclast-like giant cells. Immunohistochemically, the tumor cells were positive for vimentin, CD68 and smooth muscle actin (SMA).

Key Words : Plexiform fibrohistiocytic tumor; Neck

Plexiform fibrohistiocytic tumor (PFT) is a rare soft tissue tumor that was first described by Enzinger and Zhang in 1988, and this tumor predominantly involves the upper extremities of children and young adults.[1] Clinically, PFT presents as a painless, slowly growing subcutaneous or dermal mass. Local recurrence is common if complete excision is not achieved.[1,2] Lymph node metastasis rarely occurs,[1] and one fatal case of pulmonary metastasis has been reported.[3] Microscopically, the lesion is characterized by a multinodular or plexiform proliferation of histiocyte-like and fibroblast-like cells, and these are associated with multinucleated osteoclast-like giant cells. We herein present a rare case of PFT of the neck, and this is the second case report on this type of malady in Korea.

CASE REPORT

A 46-year-old woman who was previously healthy presented with a painless mass on the left neck that was noticed about 3 months earlier. The neck computed tomography showed an enhanced nodular lesion. Grossly, the mass was approximately $2.0 \times 1.5 \times 1.5$ cm and it was adherent to the skin. The cut surface was gray-white and firm. On low power examination, multiple small and medium-sized nodules were noted to involve the subcutaneous tissue with extension into the deep dermis and skeletal muscle (Fig. 1). The nodules were circumscribed by fibromatosis-like areas, and this created a plexiform growth pattern (Fig. 2). These nodules were composed of mononuclear histiocyte-like cells and multinucleated osteoclast-like giant cells, and sometimes there were spindle fibroblast-like cells at the periphery, and occasionally the lesions showed microhemorrhage (Fig. 3). The mononuclear histiocyte-like cells had pale cytoplasm, round-to-oval nuclei and small nucleoli, and the multinucleated osteoclast-like giant cells had abundant eosinophilic cytoplasm, three or more round nuclei, and a single prominent nucleolus. There was no cellular atypia and pleomorphism. The morphological findings were consistent with PFT. Immunohistochemically, the tumor cells were positive for vimentin, CD68 (Fig. 4) and smooth muscle actin (SMA), but they were negative for S-100 protein, CD34 and cytokeratin.

DISCUSSION

PFT is a rare mesenchymal neoplasm that usually occurs on the upper extremities of children and young adults. This tumor has a female predilection and it typically presents as a slowly growing, painless mass in the dermis and subcutaneous tissue.[1]

Only one case of this tumor being located in the scalp has been reported in Korea.[4] PFT rarely presents in the head and neck region and it is rarely found after the age of 30 years.[1,2,4] Our case is unusual in that the location of the tumor was the neck and the age of the patient was over 30 years.

Histologically, PFTs are characterized by plexiform proliferation of mononuclear histiocyte-like cells, multinucleated giant cells and spindle fibroblast-like cells in variable proportions, and the tumors have three distinct growth patterns: fibrohistiocytic, fibroblastic, and mixed, depending on the predominant cell type.[2] Our case exhibited the fibrohistiocytic pattern. PFTs do not display cellular atypia and pleomorphism. Most PFTs display fewer than 3 mitoses/10 high power fields and there are no atypical mitoses.[2] In our case, mitoses were observed infrequently, <3 mitoses/10 high power fields, and no atypical mitoses were observed. PFTs have been considered to have a low-grade malignant potential, but they have a high rate of local recurrence that varies from 35% to 40%,[1,5] and this reflects the infiltrative nature of the lesion and the difficulty in achieving a complete excision. No histologic parameters have been correlated with the more aggressive behavior.

The dual expression of CD68 and SMA is typical of PFTs.[5] The histogenesis of PFT is uncertain, but the cell of origin is proposed to be a myofibroblastic cell with the capacity for biphasic differentiation toward either a fibroblastic or histiocyte-like morphology.[5-7] Immunohistochemically, the multinucleated giant cells and the abundant mononulcear histiocyte-like cells were positive for CD68, and the spindle fibroblast-like cells and the rare mononulcear histiocyte-like cells were positive for SMA. The tumor cells were negative for S-100 protein, CD34 and cytokeratin. The results of the immunohistochemical study in our case were consistent with those of other reports.[2,5,7]

The differential diagnosis of a subcutaneous mass composed

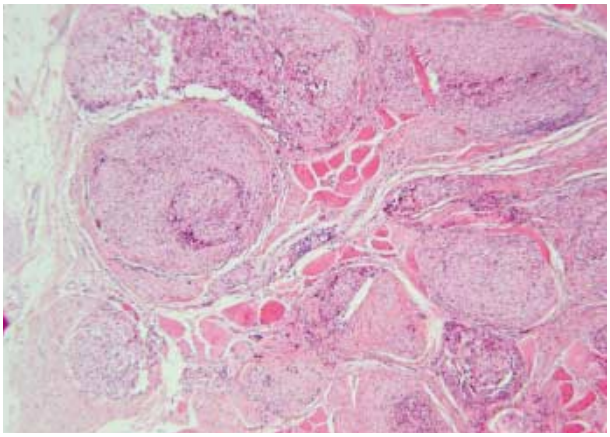

Fig. 1. The tumor consists of multiple small and medium-sized nodules that involve the subcutaneous tissue with extension into the deep dermis and skeletal muscle.

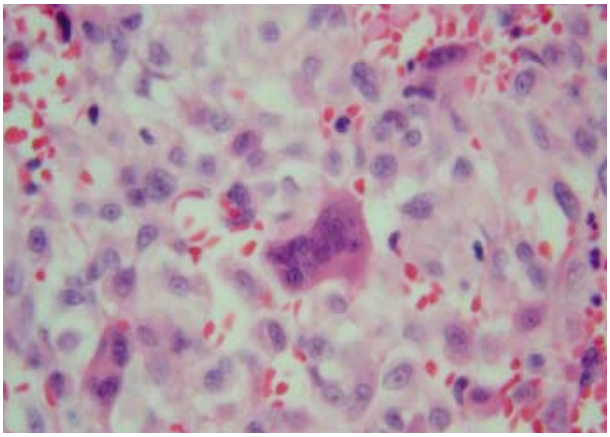

Fig. 3. The nodule is composed of mononuclear histiocyte-like cells and multinucleated osteoclast-like giant cells.

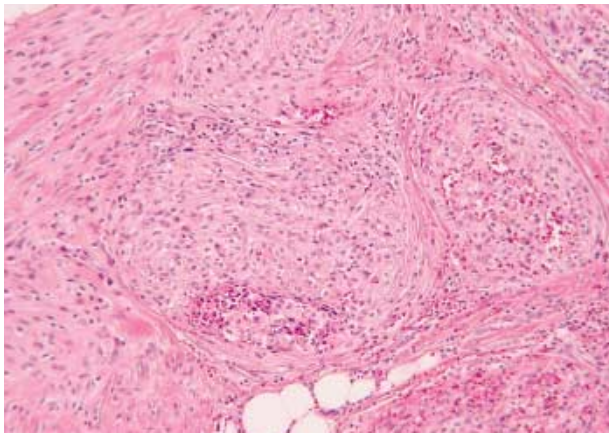

Fig. 2. The nodules are circumscribed by fibromatosis-like areas.

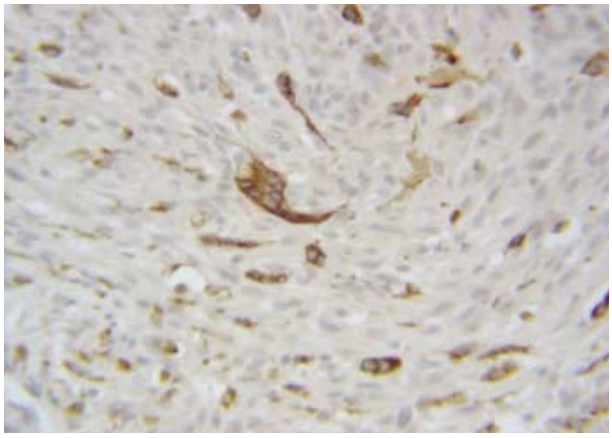

Fig. 4. The multinucleated giant cells and many of the mononulcear histiocyte-like cells are positive for CD68 by immunohistochemical staining.

predominantly of spindle cells with a plexiform growth pattern includes tuberculosis, plexiform neurofibroma, and cellular neurothekeoma. The presence of nodules made up of mononuclear histiocyte-like cells admixed with giant cells may suggest the possibility that tuberculosis should be considered in the diagnosis; however, the nodules in PFT contain osteoclast-like cells rather than Langhans-type giant cells, and the nodules do not have central necrosis. Plexiform neurofibroma was excluded because the tumor cells were completely negative for S-100 protein. Unlike PFT, cellular neurothekeoma lacks osteoclast-like giant cells and it shows frequent mitotic figures and immunohistochemical positivity for NK1/C3.

In summary, we report here on a very unusual case of PFT in the neck of a 46-year-old woman. This is the second Korean case of PFT and the first case with a cervical manifestation. Since some PFTs exhibit an aggressive behavior, careful follow-up is necessary.

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