A Case of Nerve Sheath Myxoma with Electron Microscopic Study

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We report a case of an uncommon cutaneous neoplasm of nerve sheath myxoma in a 25-year-old woman who had a papule on the left cheek of her face.

The histologic findings of the lesion showed that lobulated tumors were composed of spindle shaped and stellate tumor cells in an abundant matrix of mucosubstances. The electron microscopic findings showed numerous cytoplasmic myelin figures, and interdigitating long cytoplasmic processes. S-100 protein was not discovered and alcian blue and PAS stain showed positive.

After surgical excision of tumor, the patient did not have any evidence of disease for one year. (Ann Dermatol 6:(1)90~93, 1994)

Key Words: Electron Microscopic Findings, Nerve Sheath Myxoma

Nerve sheath myxoma was first described as a specific pathologic entity by Harkin & Reed in 1969[1] as a myxoma of nerve sheath.

After this original report, subsequent authors have offered alternative names such as bizarre cutaneous neurofibroma[2], pacinian neurofibroma[3], neurothekeoma[4], and myxoid tumor of nerve sheath[5]. It is believed that they are originated from the nerve sheath. Lesions usually occur at the ages of 2 to 30 years as a solitary nodule primarily on the face, arms and shoulders.

Recently we have encountered a case of nerve sheath myxoma of the face and studied it by using electron microscopy and immunohistochemical techniques.

REPORT OF A CASE

A 25-year-old female presented with a 6mm × 4mm sized asymptomatic, skin colored, dome shaped papule on her left cheek for 1 year period (Fig. 1).

Clinical diagnosis of granuloma pyogenicum and nevocellular nevus were suspected initially and the lesion was totally excised. Her past history and family history were noncontributary. Histological sections of excised lesions were stained with Hematoxylin-Eosin(H & E), alcian blue and periodic acid-schiff(PAS).

H & E stained sections revealed an effaced epidermis with a narrow grenz zone overlying a multilobate tumor(Fig. 2). The tumor was divided into indistinct lobules by fibrous connective tissue. Each lobule consisted of spindle or round cells lying singly or in small groups within a myxoid matrix. The mucoid matrix was positive for alcian bule(Fig. 3) and PAS statin, indicative of the presence of sulfated heteroglycans.

Immunohistochemistry, S-100 protein was negative in these cells.

Ultrastructually, the tumor cells were generally spindle shaped or irregular shaped with small and long cytoplasmic extensions which sometimes interdigitated one another(Fig. 4, 5).

Most cells contained a single nucleus that was

Received April 23, 1993
Accepted for publication June 21, 1993
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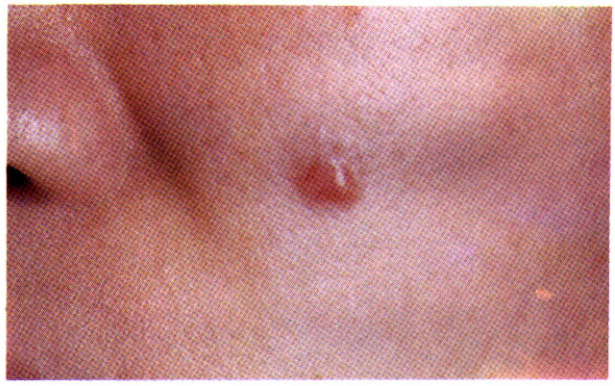

Fig. 1. A 6mm × 4mm sized, hard and non-tender papule on the left cheek.

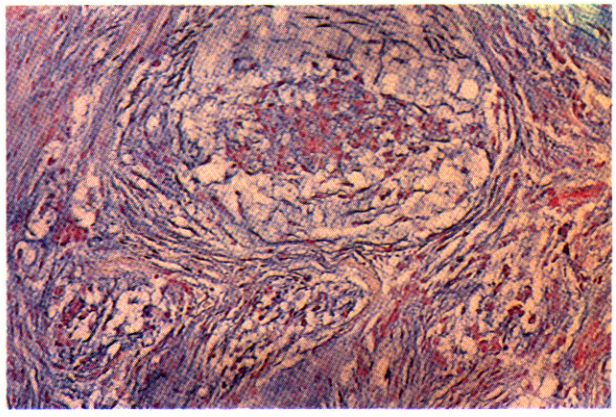

Fig. 3. The mucinous material was positively stained with alcian blue(× 400).

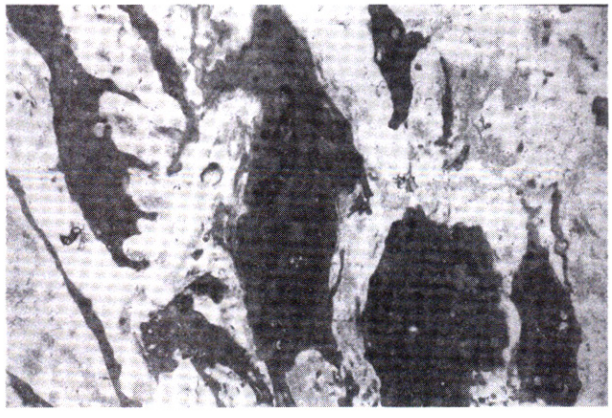

Fig. 5. Loosely arranged spindle shaped neoplasic cells with a long interdigitaing cytoplasmic extension (× 7,500).

frequently segmented or showed deep irregular indentations and pseudovacuoles(Fig. 6).

The cytoplasm contained a modest amount of organelles. Multilayered structure of concentrically arranged delicate membranes were seen in the cytoplasm of several tumor cells. Some of these membrane formations were very densely packed and markedly osmiophilic, i.e., such as the appearacnce of myelin figures(Fig. 7).

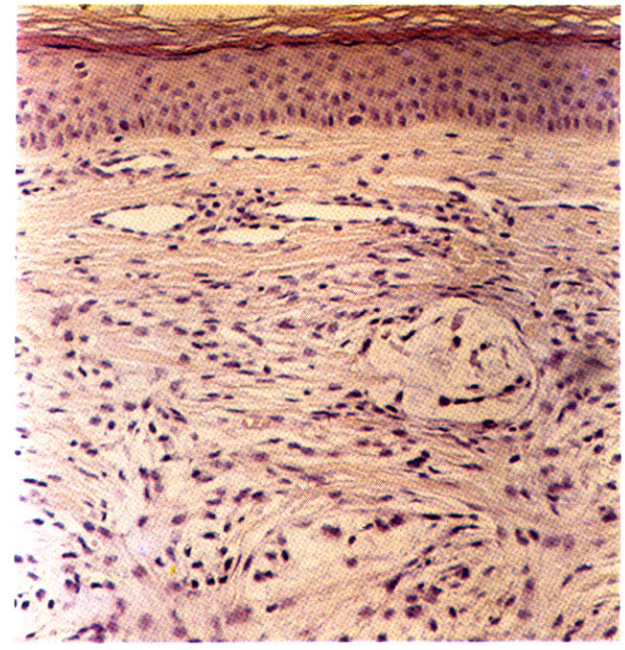

Fig. 2. Neoplastic cells in a mucinous matrix are arranged in fascicles in the reticular dermis(H & E × 40).

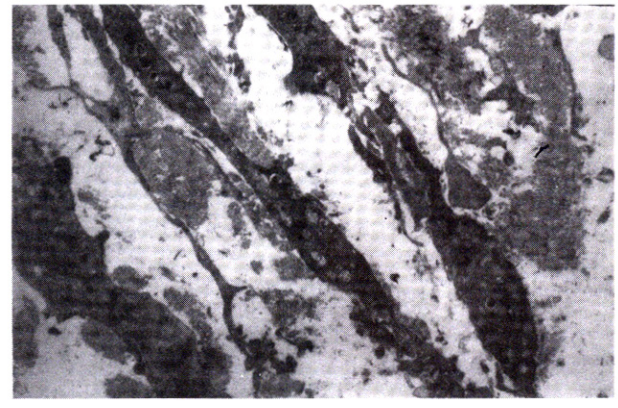

Fig. 4. Long interdigitating cytoplasmic projections between two neoplastic cells(× 30,000).

Nerve sheath myxoma was diagnosed based on clinical appearance, histologic findings and electron microscopic findings.

DISCUSSION

Nerve sheath myxoma is a rare neoplasm[6]. This

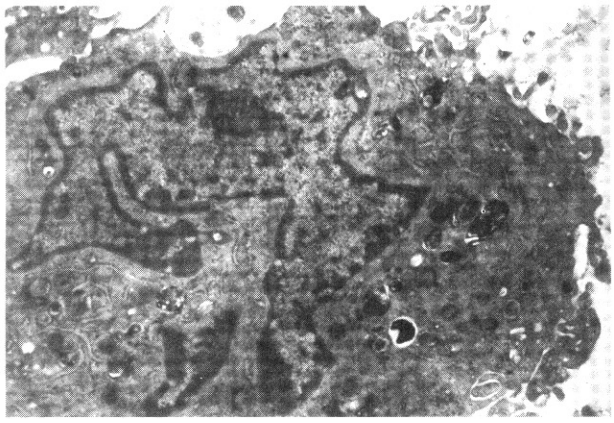

Fig. 6. A polygonal tumor cell with cytoplasmic projections. Note the indented nuclues with diffuse and marginated chromatin. Numerous lysosomal structure, residual bodies and rough endoplasmic reticulum are also present in cytoplasm(× 5,000).

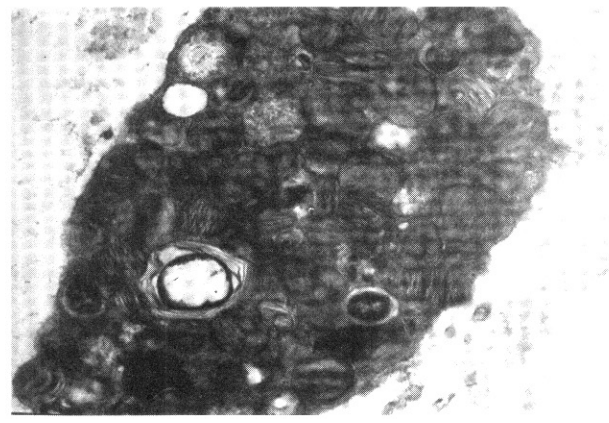

Fig. 7. A cluster of myelinoid or laminated figures in the cytoplasm of a neoplastic cell(× 13,000).

neoplams usually occurs as a solitary nodule and is measured less than 1 cm in diameter and is seen most commonly in the face but also occurs on the arms and shoulders.

The case presented of nerve sheath myxoma is light microscopically characterized by the multinodular growth of generally spindle shaped or stellate tumor cells in an abundant matrix of mucosubstance. The microscopic appearance matches very well with previous descriptions of similar tumors[1-13].

The ultrastructure of tumor cells showed prominent multilayered membrance formations of myelin figures, and long interdigitating cell processes. These findings corresponded well with peripheral nerve sheath tumors. However from the ultrastructure findings, it is not possible to state whether the tumor cells originate from Schwann cells or perineural cells.

As for the immunohistochemistry of nerve sheath myxoma, a controversy exists with regard to the presence of S-100 protein, adding further support to the concepts of the nerve sheath origin. However other reports[5,14] documented that they were unable to demonstrate S-100 protein positive. In our case, S-100 protein was not demonstrated in tumor cells. The main controversy exists as to whether nerve sheath myxoma is derived from the Schwann cells of ectodermal origin of apparent mesodermal origin. It is possible that the cells represent stages of a precursor cell of neural crest origin.

In conclusion, the microscopic, ultrastructural and immunohistochemical results of this case lead to the diagnosis of nerve sheath myxoma.

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