Neurothekeoma

— Nerve Sheath Myxoma —

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A typical case of neurothekeoma arising on the scalp, an unusual site for this tumor, is reported. The positive staining for S-100 protein suggests that neurothekeoma is a tumor of Schwann cell origin. (Ann Dermatol 2:(2) 117-120 1990)

Key Words: Neurothekeoma, Scalp

Neurothekeoma is an unusual myxoid tumor of nerve sheath origin. It was first described as a specific pathologic entity under the term 'myxoma of nerve sheath' by Harkin and Reed in 1969.[1] In 1980, Gallager and Helwig[2] reported 53 similar cases and named it neurothekeoma. The tumor appeared mostly on the face and also on the extremities and trunk but very rarely on the scalp.[3] We report a case of this tumor on the scalp in a 44-year-old female patient.

REPORT OF A CASE

A 44-year-old woman visited our hospital in April 1989, with a mass on the scalp present for 28 years. The past and family history were noncontributory. She had had this mass for the past 28 years without change, but recently the lesion increased in size. Cerebriform pedunculated,

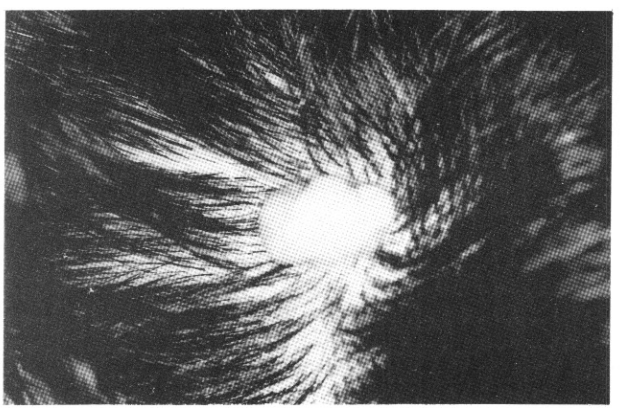

Fig. 1. A 1.5×0.8cm, flesh colored, cerebriform, pedunculated nodule on the scalp.

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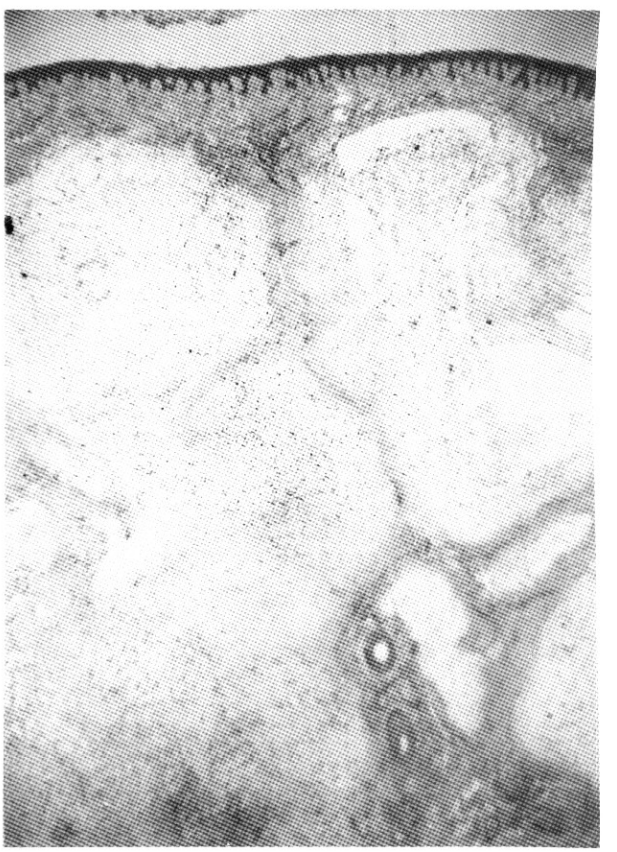

Fig. 2. The tumor is situated entirely in the dermis and the superficial panniculus. There is a well defined grenz zone and is no membrane or capsule. (H & E stain, ×20).

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tender nodule on the scalp (Fig. 1). There was occasional bleeding with combing. Physical examination showed a 1.5×0.8cm, flesh-colored. The mass was removed surgically.

Histopathologically, there were no changes in the epidermis except for elongation of the rete ridges. The tumor was situated entirely in the dermis and superficial panniculus. There was a grenz zone and it was well defined with no membrane or capsule (Fig. 2). There was a mild to moderate infiltrate of mononuclear cells and plasma cells and telangiectasia in the fibrous stroma. It was divided into multiple distinct cellular nests by thickened fibrous connective tissue (Fig. 3). The nests were variable in size and oval to irregular shaped (Fig. 4). Each nest consisted mostly of spindle to oval shaped cells lying singly or in small groups with abundant mucinous background (Fig. 5). Bizarre cells with hyperchromatic nuclei were often observed, however mitotic figures were absent. The myxoid stroma stained positive with alcian blue.

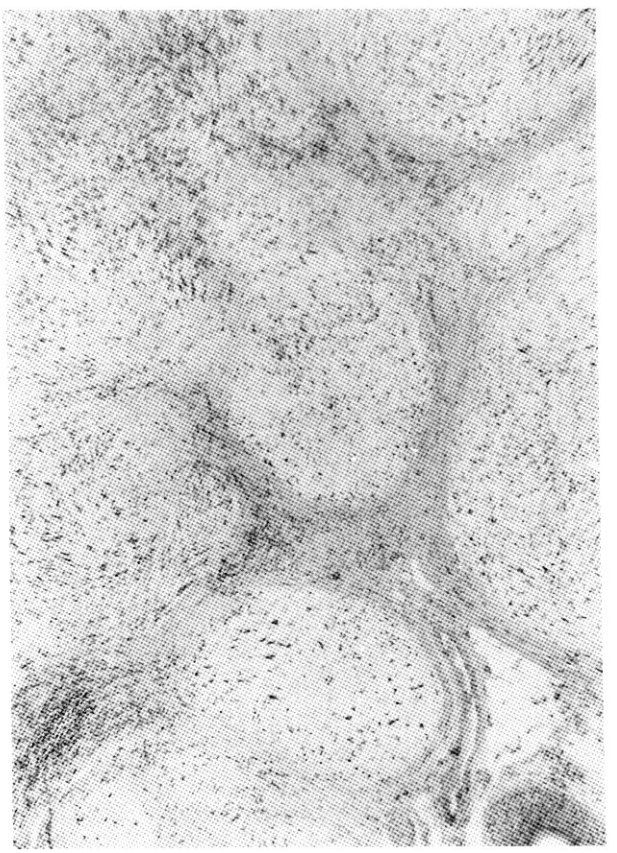

Fig. 3. The tumor is divided into multiple distinct nests by thickened fibrous connective tissue. (H & E stain, ×40).

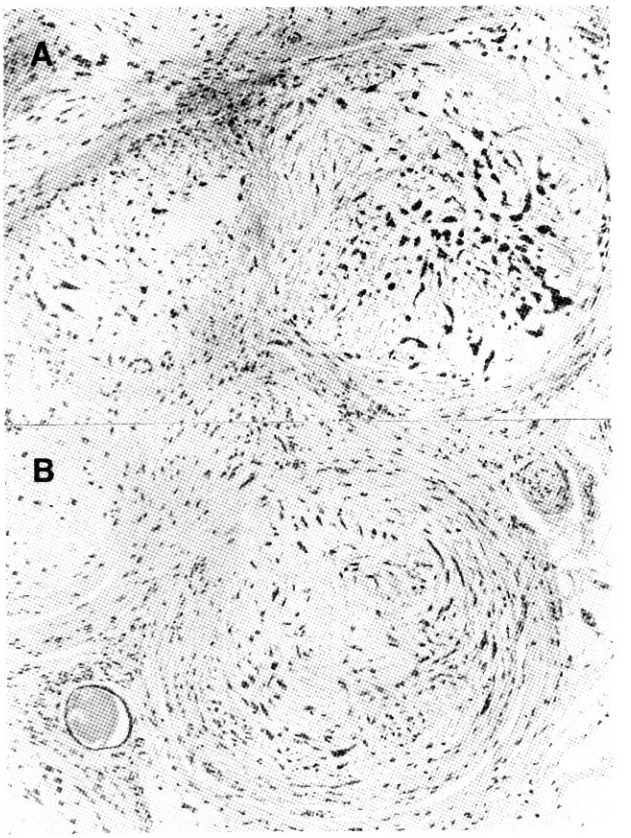

Fig. 4. The nests are irregular shaped and variable sized (A and B; H & E stain, ×100).

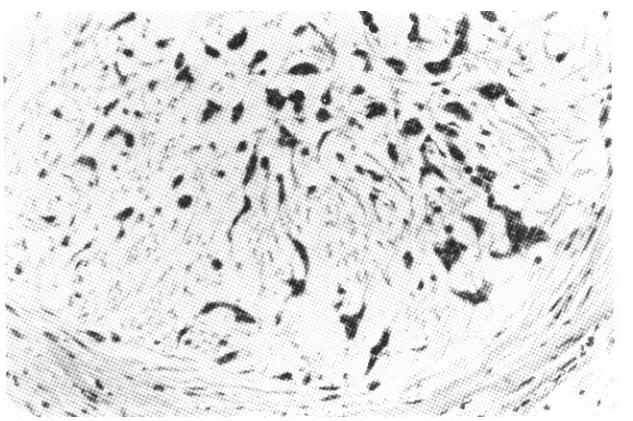

Fig. 5. Each nest consisted mostly of spindle to oval shaped cells lying singly or in small groups with an abundant mucinous background (H&E, ×400).

Using an immunohistochemical stain for S-100 protein, the nuclei or cytoplasm of the spindle, stellate and epithelioid tumor cells were stained uniformly strongly positive in myxomatous areas (Fig. 6). Many tumor cells also revealed a less prominent staining of the cytoplasm. In the cross sec-

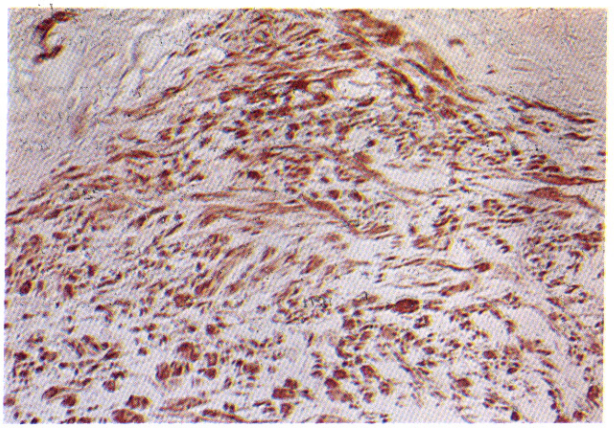

Fig. 6. The nuclei of the tumor cells stained uniformly for S-100 protein (Immunoperoxidase stain, ×400)

tion of a peripheral nerve, the nerve fascicles were stained, but the epineurium, which is the outer layer of the peripheral nerve did not take up the stain. Overlying epidermal melanocytes and Langerhans cells also were stained as were the secretory cells of the eccrine sweat gland.

DISCUSSION

Since this benign tumor of nerve sheath origin was first described as a myxoma of the nerve sheath by Harkin and Reed[1] in 1969, various terms have been used for this tumor such as: neurothekeoma,[2] nerve sheath myxoma,[3] bizarre cutaneous neurofibroma,[4] and myxoid tumor of nerve sheath.[5]

Neurothekeoma is a term to stress its nerve sheath origin (from the Greek Oeke, meaning sheath).[2] These tumors usually arise during childhood and early adult life. Females are affected nearly twice as often as males.The lesions appear as nodules of flesh color on their cut surface and they average one centimeter in size. The face is the most common site, accounting for almost one-third of the lesions and it also occurrs on the extremities and trunk.[3] Among the 40 cases reported by Gallager and Helwig[2] there were none located on the scalp. Among 64 cases reported by Pulitzer & Reed (3), only 4 cases (6%) were found to be on the scalp. Thus, our patient's scalp tumor was in an unusual location. This tumor is invariably benign. Only one of 70 lesions recurred and no metastses are known to have occurred.[3]

Histologically, the lesion involves the reticular dermis and the superficial portion of the subcutis. In most examples, the papillary dermis is spared. It is nodular and well delineated but not encapsulated. The tumor is separated into distinct lobules by fibrous connective tissue that has been compared to the perineurium. Each lobule consists of a background of myxomatous tissue with few cells and abundant pale matrix material. The cells are minimally pleomorphic, benign appearing, stellate, elongated, and bipolar in shape. Many cells are markedly hyperchromatic and show moderate atypia, and at most only rare mitotic figures. The mucinous background matrix within the cell nests has been demonstrated to be acid mucopolysaccharides, usually hyaluronic acid. Also, there is a significant component of sulfated acid mucopolysaccharides, presumably chondroitin sulfate.[6] Neoplastic perineurial and Schwann cells have the capacity to produce hyaluronic acid and sulfated glucosaminoglycans.[6,7]

Electron microscopically, the tumor cells are compatible with Schwann cells[2,8] or perineurial cells.[3] However, Schwann cells can not be distinguished from neuroectodermal perineurial cells by any of the present techniques including histochemistry, electron microscopy, or tissue culture.[9] It is not known if the perineurial cells contain S-100 protein.[7,10]

Immunoperoxidase staining for S-100 protein is often positive in tumor cells of neurothekeoma as in our case.[3,7,8] S-100 protein is a neuroectodermal marker of the Schwann cell melanocytic tumors.[11] The above immunohistochemical study suggests that neurothekeoma is a tumor of Schwann cell origin.

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