

Dermal Nerve Sheath Myxoma

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Citation

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Abstract

We report a rare case of a dermal nerve sheath myxoma of the left intranasal skin occurring in a 33-year-old female. A brief review of nerve sheath myxoma and a similar tumor, neurothekeoma, is presented.

CASE REPORT

A 33-year-old female presented with a 5-month history of a left intranasal mass. A lesion from the same location was biopsied previously that was interpreted as a myxoid fatty tumor. The enlarging mass was surgically excised.

On microscopic examination, the tumor was located in the dermis. It had a multilobular architecture with abundant myxoid matrix surrounded by a fibrous pseudocapsule. The lobules were composed of stellate, spindle cells with small spindle or ovoid nuclei [Fig 1, 2]. Foci of increased cellularity were noted [Fig 3]. Overall, the cells were bland with no increased mitotic figures and there was no tumor necrosis. The tumor cells were positive for Vimentin [Fig 4] and S-100 protein [Fig 5] immunochemical stains. A diagnosis of a dermal nerve sheath myxoma was made.

Figure 1

Figure 1: Dermal nerve sheath myxoma, low power view.

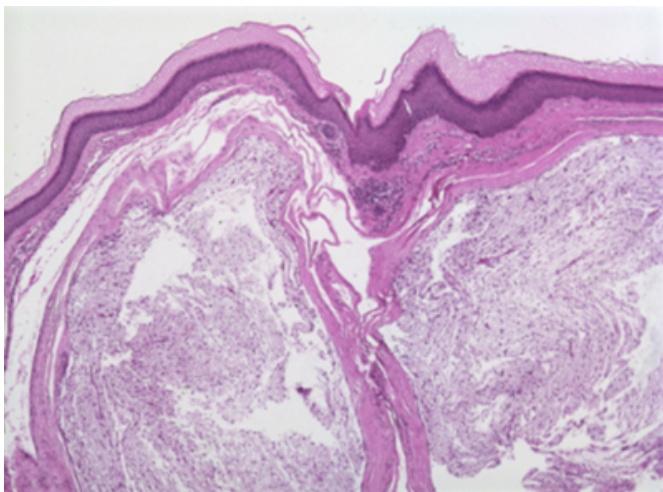


Figure 2

Figure 2: Dermal nerve sheath myxoma, H&E

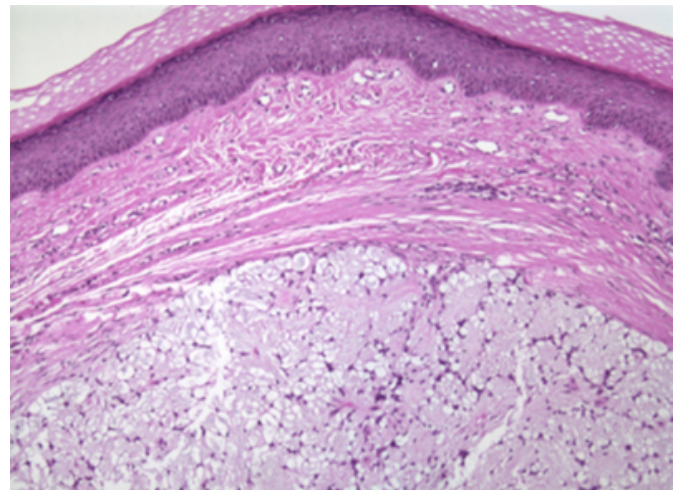


Figure 3

Figure 3: Dermal nerve sheath myxoma, high power view.

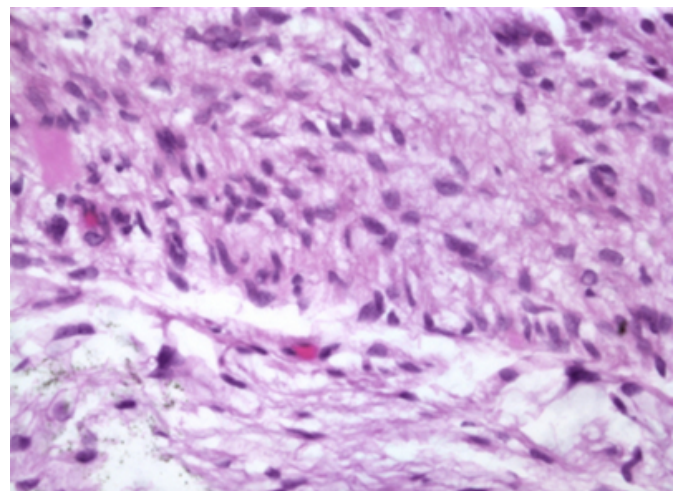


Figure 4

Figure 4: Dermal nerve sheath myxoma, positive Vimentin stain

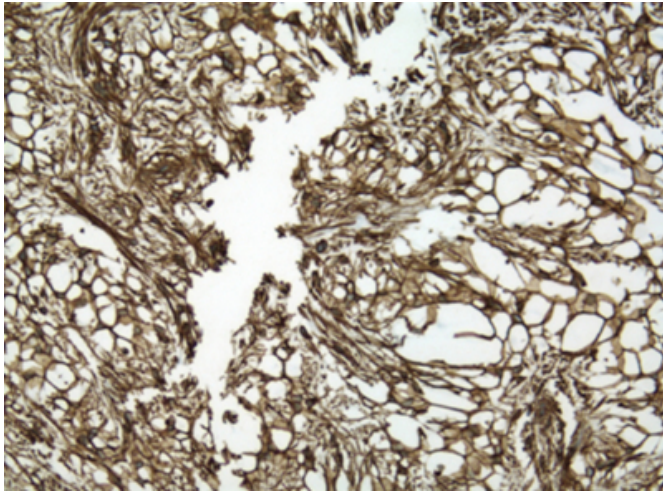
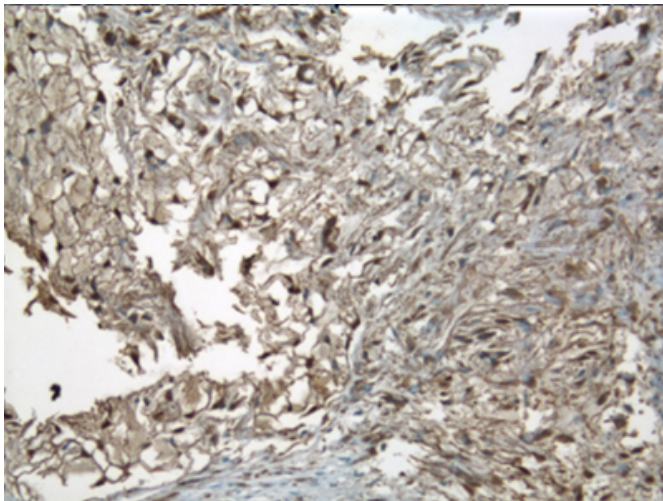


Figure 5

Figure 5: Dermal nerve sheath myxoma, positive S-100 protein stain



COMMENT

Nerve sheath myxoma is a rare tumor of nerve sheath origin which typically affects young adults, with a predilection for females (2:1). This soft tissue tumor was first described by Harkin and Reed in 1969^[1]. The tumor was further studied in greater detail by Pulitzer in 1985^[2]. Nerve sheath myxoma has also been referred to as cutaneous lobular neuromyxoma, perineurial myxoma, and pacinian neurofibroma.

Nerve sheath myxoma is a superficial, multilobulated, predominantly myxoid, spindle cell neoplasm exhibiting Schwann cell differentiation. The typical lesion presents as a firm slow-growing painless nodule. The tumor mostly

occurs in the upper limbs or head and neck area. It usually involves the reticular dermis and often extends into the subcutaneous fat. Lobules, usually separated by fibrous septae, are composed of an abundant myxoid matrix containing spindle cells arranged in fascicles and whorls. Cells with a more epithelioid appearance are also often encountered. Mild nuclear pleomorphism and rare mitoses may be present. Focally, there may be nuclear atypia and normal mitotic figures. Immunohistochemically, nerve sheath myxoma is strongly positive for S-100 protein, GFAP, and Vimentin. Nerve sheath myxoma is a benign neoplasm cured by simple excision. However, a high local recurrence rate has been documented ^[4].

Many pathologists and dermatologists believe that nerve sheath myxoma is a subtype of neurothekeoma (so-called myxoid, hypocellular variant of neurothekeoma). The clinical appearance of nerve sheath myxoma is indistinguishable from that of a neurothekeoma. Both tumors have a similar size ranging from 0.5 to 3.0cm and are solitary, well-circumscribed, and sometimes multilobular.

Microscopically, both tumors are multinodular with a variable amount of myxoid stroma. However, recent ultrastructural and immunohistochemical studies ^[3,4] indicate that they are two distinct entities. Nerve sheath myxoma shows ultrastructural features of Schwann cell differentiation and S-100 protein immunoreactivity. Neurothekeoma exhibits features of fibroblasts (CD34 positive) or undifferentiated stromal cells. It also shows evidence of smooth muscle differentiation and negative staining for S-100 protein.

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