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Case Report:

Myxoid Neurothekeoma of the Nipple.

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Abstract: Neurothekeomas are rare benign cutaneous neoplasms of nerve sheath origin. They are primarily found in the superficial soft tissue and are also known as dermal nerve sheath myxomas. They are commonly found on the upper extremities, head and neck followed by trunk. Here is an unusual presentation of neurothekeoma occurring as a polypoidal lesion of the nipple in a young female patient.

Key Words: Neurothekeoma; Dermal nerve sheath myxoma

Introduction:

Neurothekeomas or dermal nerve sheath myxomas are slow growing tumors of nerve sheath origin. They are common on the upper extremities, head and neck. We are reporting an unusual presentation of neurothekeoma as a polypoidal mass over the nipple, clinically mimicking a nipple papilloma.

Case Report:

A 26 years old south Indian woman presented with a slow growing, non-tender, polypoidal lesion over the left nipple for 3 years. There was no associated nipple discharge or lump in the breast. A clinical diagnosis of nipple papilloma was made; the lesion was excised completely and sent for histopathological examination.

The excised specimen was a skin covered polypoidal tissue, measuring 2 cm in greatest dimension with a stalk measuring 0.5 cm; the cut surface was grey-white, lobulated and glistening (Fig 1). Microscopic examination revealed a lobular, hypocellular neoplasm in the dermis (Fig 2). Bland spindle shaped tumor cells with interspersed collagen bundles were seen embedded in a myxoid stroma (Fig 3). Nuclear pleomorphism and mitotic figures were absent. Immunohistochemistry revealed strong S-100 positivity of the tumor cells (Fig 4). EMA, CD34 and HMB-45 were negative. The surgical margins were completely free of tumor. With the above features, a diagnosis of myxoid neurothekeoma was given.

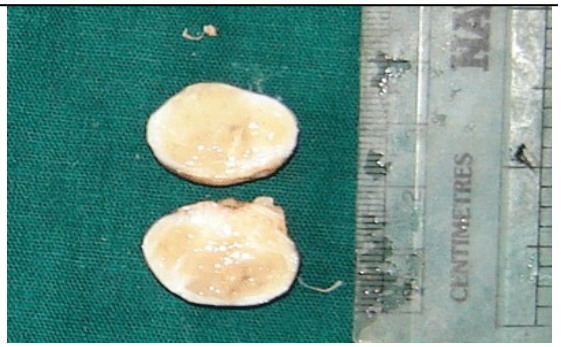

Figure 1: Photograph showing skin covered polypoidal tissue with a grey white glistening surface

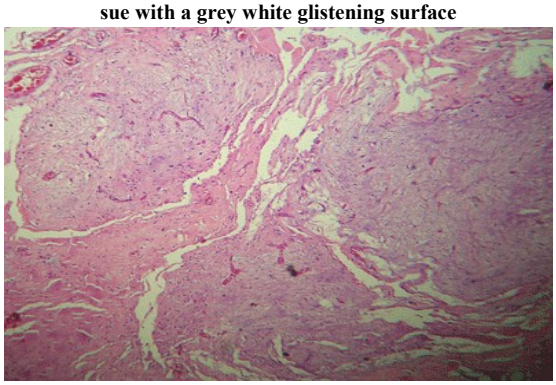

Figure 2: Photomicrograph showing a lobular, hypocellular neoplasm in the dermis (x100)

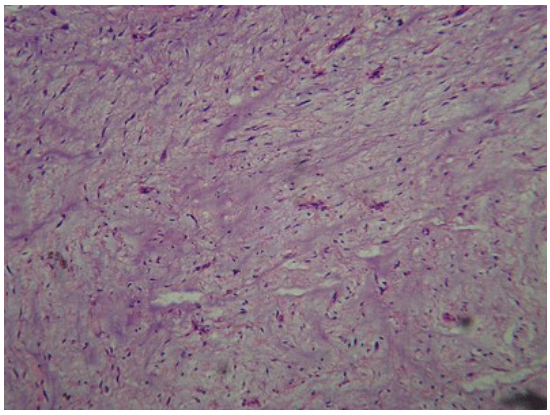

Figure 3: Photomicrograph showing abundant myxoid stroma containing spindle shaped tumor cells (x100)

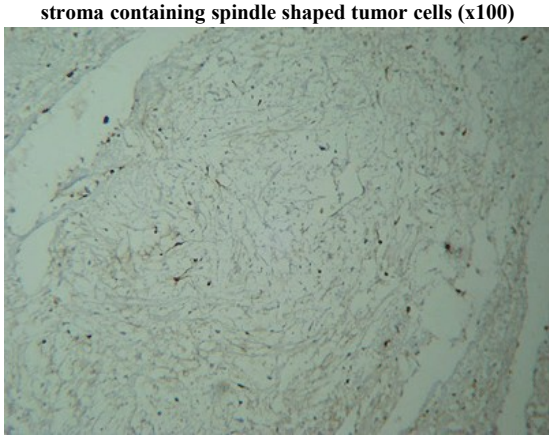

Figure 4: Photomicrograph showing strong S-100 positivity of the tumor cells (x100)

Discussion:

In the year 1969, Harkin and Reed first described a rare neoplasm arising in the endoneurium of peripheral nerves, characterized by abundant myxoid matrix and called it myxoma of nerve sheath.(1) The term neurothekeoma was coined by Gallager and Helwig who first published a large series of this tumor in 1980. Neurothekeoma was described in detail by Pulitzer and Reed in 1985.(1) Females in the 2nd and 3rd decades of their lives were more commonly affected with rare occurrence in infants and elderly. These tumors showed prediction to head and neck, arms, shoulders and trunk; other uncommon sites of involvement were subungal region, eye and spine. Out of the 300 cases of neurothekeoma described by Pulitzer and Reed and Gallager and Helwig, only one case had the lesion over the breast. Ours is the first reported case of neurothekeoma arising over nipple and presenting as a polypoidal growth.

In 1986, Rosati described a similar tumor with high Cellularity and named it cellular neurothekeoma. Fetsch and others reported that cellular neurothekeoma exhibited different immunohistochemical profile and occurred in younger patients with a predilection to head when compared to myxoid neurothekeomas. 3 distinct types of neurothekeoma were described (2):

a. Hypocellular type (myxoid): The hypo cellular group consisted of well circumscribed lobular tumors with prominent myxoid stroma and positive for S-100 and collagen type IV and variably positive for EMA.

b. Cellular type: The cellular groups were composed of ill defined nests and fascicles of spindle cells with scant mucin and S-100 negativity. The cellular neurothekeomas do not possess any evidence of neural differentiation and therefore represented cutaneous neoplasm of undetermined cellular origin.

c. Mixed type: Variable Cellularity and mucin content with poor demarcation and variable results with immunomarkers.

However, Colonje et al showed that the cellular neurothekeoma was negative to PGP 9.5 and strongly positive for NK/C3. Thus they proposed that cellular neurothekeoma represented epithelioid variant of pilar leiomyoma.(3)

Laskin et al in 2000 showed that myxoid/ hypocellular variety occurred more commonly in male patients with a peak incidence in fourth decade and were found in both upper and lower limbs and back.(4) This was in contrast to cellular neurothekeoma which affected more female patients with peak incidence in second decade of life in upper body distribution.

The fact that tumor location changes with age was shown by Papadopoulos et al. In children, head and neck comprised 45.5% of cases as compared to 24.4% in adults. In adults, upper extremity tumors were more common.(5) Hornick et al in their study of 133 cellular neurothekeomas, showed that 35% arose in upper extremities, 33% in head and neck, 17% in lower limb and 15% on trunk; of all these cases, face and shoulder were the most commonly affected sites.(6)

The tumors range from 0.4 to 4.5 cm in greatest dimension, with a rubbery to firm consistency, and on cut section, small, well-demarcated, translucent or whitish (rarely yellowish), glistening, mucoid nodules are often noted.(7)

In our patient, histology showed a well circumscribed, multilobulated tumor in the dermis, composed of bland stellate and spindle cells dispersed in abundant myxoid stroma. Further, immunohistochemistry demonstrated S-100 positivity of the tumor cells, confirming the neural origin of this tumor.

The tumors are treated by wide local excision and surgical clearance as local invasion and tumor recurrence rate is high.(8)

To our knowledge, this is the first case report of a myxoid neurothekeoma involving the nipple and presenting clinically as a Polypoidal mass.

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