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)
a. Dilection to head when compared to myxoid neurothekeomas. 3
b. Tochemical profile and occurred in younger patients with a pre
and named it cellular neurothekeoma. Fetsch and others repor
over nipple and presenting as a polypoidal growth.

Discussion:

In the year 1969, Harkin and Reed first described a rare neo-
plasm arising in the endoneurium of peripheral nerves, charac-
terized by abundant myxoid matrix and called it myxoma of
nerve sheath.(1) The term neurothekeoma was coined by Gall-
ger 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 in-
fants and elderly. These tumors showed predilection to head and
neck, arms, shoulders and trunk; other uncommon sites of in-
volvement 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 repor-
ted that cellular neurothekeoma exhibited different immunohis-
tochemical profile and occurred in younger patients with a pre-
dilection to head when compared to myxoid neurothekeomas. 3
distinct types of neurothekeoma were described (2):

a. Hypocellular type (myxoid): The hypo cellular group con-
sisted of well circumscribed lobular tumors with promi-
inent 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 mu-
cin and S-100 negativity. The cellular neurothekeomas do
not possess any evidence of neural differentiation and
therefore represented cutaneous neoplasm of undeter-
mined cellular origin.

c. Mixed type: Variable Cellularity and mucin content with
poor demarcation and variable results with immunomark-
ers.

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 epithel-
oid variant of pilar leiomyoma.(3)

Laskin et al in 2000 showed that myxoid/ hypocellular variety
occurred more commonly in male patients with a peak incid-
ence in fourth decade and were found in both upper and lower
limbs and back.(4) This was in contrast to cellular neur-
othekeoma which affected more female patients with peak in-
cidence 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),
glistenning, mucoid nodules are often noted.(7)

In our patient, histology showed a well circumscribed, mul-
tilobulated tumor in the dermis, composed of bland stellate and
spindle cells dispersed in abundant myxoid stroma. Further, im-
munohistochemistry demonstrated S-100 positivity of the tu-
more 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 neur-
othekeoma involving the nipple and presenting clinically as a
Polyoidal mass.

References:

Berkere DAR. Subungal neurothekeoma. J Am Acad Dermo-
2. Fetsch JF, Laskin WB, Hallman JR, Lepton GP, Miettin-
en. Neurothekeoma – An analysis of 178 tumors with de-
tailed immunohistochemistry data and long term patient
‘neurothekeoma’ an epithelioid variant of pilar leiomyoma? Mor-
4. Laskin WB, Fetsch JF, MiettinenM. The “neur-
othekeoma”: immunohistochemical analysis distinguishes
the true nerve sheath myxoma from its mimics. Hum Pathol.
2000;31:1230-1241.
5. Papadopoulos EJ, Cohen PR, Hebert AA. Neurothekeoma
– report of a case in an infant and review of the literat-
6. Hornick JL, Fletcher CDM. Cellular neurothekeoma: De-
tailed characterization in a series of 133 cases. Am J Surg
7. Argenyi ZB, LeBoit PE, Santa Cruz D, Swanson PE,
Kutzner H. Nerve sheath myxoma (neurothekeoma) of
skin: light microscopic and immunohistochemical re-
8. Ward JL, Prieto VG, Joseph A, Chevray P, Kronowitz S,
Sturgis EM. Neurothekeoma. Otolarynol Head Neck