Epithelial Sheath Neuroma with Extension to the Subcutis

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Intro

Epithelial sheath neuroma (ESN) was first described in 2000 by Requena et al (1) as a neoplasm that clinically presents as an erythematous papule or nodule on the mid-to-upper back of middle-aged to elderly adults. ESN has been reported to be asymptomatic in some cases, while exhibiting pruritus or pain in others. (2) Histologically, ESN is described to occur in the superficial dermis and is categorized by squamous epithelium surrounding enlarged peripheral nerve fiber proliferations. (3) Zelger and Zelger (4) theorized that ESN may be a reactive process, and Hirano-Ali et al (5) were the first to report a case of ESN with a direct connection to the epidermis. All reported cases have been noted to be cured by shave biopsy and/or simple excision. (1-6)

Case Report

An 82-year old woman presented with a new onset 1.2 x 1.2 cm painful nodule on her right mid-back which she stated had been present for 3 weeks. The pain was described as a near constant burning and aching sensation which was made worse with any contact, including light touch via clothing. The lesion was not present 9 months earlier during a routine dermatologic examination. The patient had no prior history of surgery or trauma to the site of the lesion. The clinical impression was basal cell carcinoma versus squamous cell carcinoma and a shave biopsy was performed.

Shave biopsy showed fascicles of spindled cells in the superficial reticular dermis with features characteristic of enlarged nerve bundles, each encircled by cytologically bland keratinocytes (fig 1a). The neural origin of the spindled cells was confirmed with S100 immunoperoxidase analysis (fig 1b).

The patients pain continued subsequent to the biopsy for 3 weeks, at which point wide local excision extending down to the level of deep fat was performed for symptom control. Histological examination of the excision showed a sizeable portion of skin extending into the subcutis. There were residual enlarged nerve bundles girdled by keratinocytes in the superficial dermis adjacent to the prior biopsy site (fig 2a). Interestingly, a few such bundles with encompassing keratinocytes also extended well into the subcutis (fig 2b and fig 2c).

After the excision, the patient became pain free over the course of days. The patient has followed up multiple times over 12 months and continued to be pain free with no recurrence of the lesion to date.

Discussion

ESN is a very rare entity with 9 reported cases to date at the time of this writing. ESN is characterized clinically as a red papule or nodule, which may or may not be symptomatic, located on the back of middle-aged to elderly adults. Histologically, ESN presents as bundles of enlarged peripheral nerve sheaths surrounded by uniform squamous epithelium often bearing a plaque-like extension in the superficial dermis. (1) Some cases exhibit an associated fibroplasia with mucin deposition around the bundles. (2) Focal dyskeratosis may be present within the epithelial sheath. Scattered lymphocytes and/or plasma cells have also been described in ESN. The differential diagnoses of greatest practical importance are perineural invasion of an invasive well differentiated squamous cell carcinoma, as well as pseudo-perineural invasion sometimes seen at surgical sites. (7) A traumatic neuroma with entrapped epithelial structures is also a consideration. The lack of scar, presence of enlarged nerve fibers in the superficial dermis, and clinical history make ESN a unique entity. (2)
The pathogenesis of ESN has not been entirely elucidated. Some have postulated a neoplastic etiology or hamartoma with late clinical onset, while more recent articles have supported a reactive process. (1,4,5) If ESN is indeed a reactive process, the underlying pathogenesis may be related to rubbing or an alternative external stimulus leading to an inflammatory response inducing various cytokines which ultimately lead to nerve hyperplasia. (4,5) In addition, the reactive enlarged nerves may be stimulating keratinocyte proliferation around them via the release of interleukin-6. (7)

Hirano-Ali et al (5) described a case in a patient with papular urticaria showing a connection to the epidermis and pointed out that there have been several cases of ESN showing overlying epidermal change consistent with lichen simplex chronicus. Moreover, Vandergriff et al (6) reported a case of multifocal ESN with two papules clinically 1 cm apart with an associated acanthotic epidermis on pathology. It is possible that these were 2 separate lesions, but also reasonable to think they could be one in the same arising as part of a reactive process secondary to the irregularity in distribution of the nerve fibers and stroma characteristic of ESN. ESN may be a combined reactive process with both enlarged nerve fibers and surrounding epithelium secondary to an external trigger.

Each case of ESN reported has been described to be cured with shave biopsy or simple excision. (1-6) We report the first case of ESN to extend down to the subcutis, raising the possibility that not all cases of ESN are confined simply to the superficial dermis. There have been cases of ESN reported to be associated with dermal cysts, and some have theorized that traumatic implantation could play a role in some or all cases of ESN. (7,8) Of note, the histological features in our case, with no reactive changes in the subcutis around the ESN residuum, argue against traumatic implantation via shave biopsy as the cause of subcutal involvement.

Due to the rarity of ESN, it is not known what percentage of cases may extend deeper than previously thought. In addition, given the extension into the subcutis, a more superficial excision may not be adequate, particularly if the patient presents with distressing symptoms. Based on our case, an excision with wide margins and down to the level of deep fat may be considered in all cases that do not clearly show a demarcation on shave or excisional biopsy. Alternatively, Mohs surgery may be considered as an alternative treatment option in attempt to adequately clear ESN margins.

References:


Figures:

Figure 1:
Thickened nerve bundles surrounded by a nest of cytologically bland keratinocytes, in the superficial dermis (fig 1a). S100 immunoperoxidase analysis highlights the enlarged nerve bundles (fig 1b).
Figure 2:

Excision specimen with residual epithelial sheath neuroma adjacent to the prior biopsy site (fig 2a), and also extending into the subcutis (fig 2b and 2c).