

## CASE REPORT

# Perineurioma of scalp in an infant: A case report with short review of literature

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## ABSTRACT

Perineuriomas are rare benign peripheral nerve sheath tumor composed exclusively of perineurial cells. They have been classified into two main types according to their location – intraneural and extraneural (soft-tissue). Extraneural perineurioma are uncommon, found mostly in the soft tissue of the extremities and trunk, rare at other site. They mainly affect adults and have been sometimes described in children but are extremely rare in infants. We present a case of extraneural (soft-tissue) perineurioma of scalp in a 3-month-old infant, highlighting clinical, pathologic, and immunohistochemical features along with a discussion of the main differential diagnosis of this tumor. This is also the youngest case ever reported in the literature.

**Key words:** Extraneural perineurioma, infant, scalp swelling

## Introduction

Perineuriomas are uncommon benign peripheral nerve sheath tumors exclusively composed of well-differentiated neoplastic perineurial cells.<sup>[1]</sup> They are classified into two main types: intraneural and extraneural (soft-tissue). Intraneural perineuriomas are restricted to major peripheral nerve boundaries, while extraneural perineuriomas are found mainly in soft tissues and skin and usually have no connection to peripheral nerves.<sup>[2]</sup>

Extraneural perineurioma usually present as a small, well circumscribed, firm, and painless mass. They mainly affect adults and few cases have been described in children, but are very rare in infants. It occurs most commonly in the soft tissue of the extremities and the trunk, infrequently in other locations.

We report a case of extraneural (soft-tissue) perineurioma occurring on the scalp of a 3-month-old infant.

## Case Report

A 3-month-old female infant was brought by her parents to the outpatient Department of Pediatric Surgery with scalp swelling. The swelling developed after birth and was gradually increasing in size. On physical examination, it was a well circumscribed, firm and nontender mass over the scalp [Figure 1a].

When she was 6-month-old, surgery was planned and the specimen was sent to histopathology section.

Grossly, a dome shaped gray white soft-tissue piece measuring 6.5 cm × 4 cm × 1.5 cm was received. Cut surface was homogenous, gray white, smooth, and glistening [Figure 1b].

Microscopically, there was a encapsulated proliferation of bland spindle cells arranged in vague storiform pattern and also forming whorls. At some places, there was formation of tiny “onion bulbs” [Figure 2]. Based on light microscopic findings, a probable diagnosis of perineurioma was made which was confirmed on immunohistochemistry. The tumor cells showed positivity for epithelial membrane antigen (EMA) but were negative for S100 and smooth muscle actin (SMA) [Figure 3].

The follow-up of the patient has been unremarkable so far.

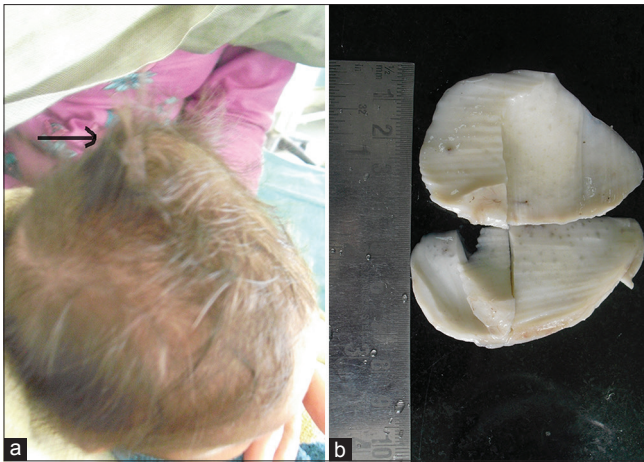
## Discussion

Perineurioma is composed of perineurial cells. Normally, these cells constitute the perineurium function as a protective perifascicular diffusion barrier situated between the epineurium and endoneurium layers of the nerves that surround both myelinated and unmyelinated peripheral nerve fascicles.

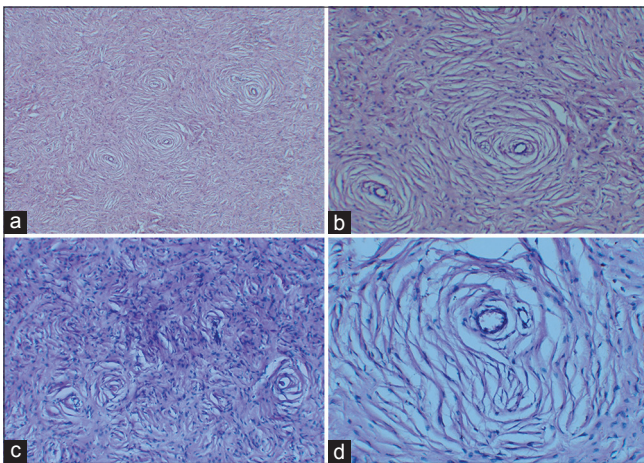
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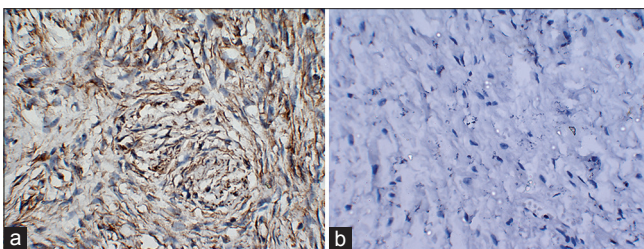
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**Figure 1:** (a) Clinical photograph of the infant with scalp swelling. (b) Gross photograph of the excised swelling revealing smooth glistening cut surface



**Figure 2:** Microscopic photograph. (a-c) The tumor cells are arranged in vague storiform pattern and forming whorls (H and E,  $\times 40$ , H and E,  $\times 100$ , H and E,  $\times 100$ ). (d) Onion bulb (H and E,  $\times 200$ )



**Figure 3:** Immunohistochemistry photograph (a) epithelial membrane antigen positive (b) S100 negative

Perineural cells ensheathing nerve fascicles are present in several tumors such as nerve sheath myxoma, traumatic neuroma, and neurofibroma. However, the term perineurioma is used strictly in which tumors formed purely of perineurial cells. They are uncommon and poorly recognized.<sup>[3]</sup>

They were first described by Lazarus and Trombetta in 1975 on the basis of ultrastructural finding. According to them,

ultrastructural features of perineurial cells include long thin cytoplasmic processes with numerous pinocytotic vesicles, abundant collagenous stroma, in-continuous basal lamina, and rudimentary intercellular junctions.<sup>[4]</sup> Since then, electron microscopy has been considered the gold standard technique in confirming the diagnosis of perineurioma. Further, in 1985 Pinkus and Kurtin first described EMA expression by immunohistochemistry in perineurial cells.<sup>[5]</sup>

Extraneural (soft-tissue) perineurioma is a benign peripheral nerve sheath neoplasm, commonly found in the subcutaneous tissues of the trunk and extremities and more often in the superficial soft-tissue (70%) than in deep tissues. They usually present as a painless, solitary, firm mass, generally not associated with an identifiable nerve as is in this case.<sup>[6]</sup>

Few cases have been described in the head and neck area, stomach, retroperitoneum, brain, kidney, maxillary sinus, mandible, and intestines.<sup>[7]</sup> They are seen in young to middle-aged adults and has no sex predilection.<sup>[8]</sup> Our case is a 3-month-old infant with perineurioma of the scalp and is the youngest case so far reported in the literature.

Macroscopically, the tumors are usually gray to white, well circumscribed but may or may be encapsulated, firm in consistency, and size varies from 0.3 to 20 cm. Histologically, extraneural perineurioma are composed of elongated spindle-shaped neoplastic cells, wavy-shaped nuclei with tapering ends and elongated, thin cytoplasmic processes forming whorls, lamellar or storiform arrangement. Their cytoplasm is slightly eosinophilic with indistinct cell boundaries. The stroma may be myxoid, collagenous, or hyalinized. Mitotic figures and necrosis are usually absent.

The immunohistochemical findings are required for accurate diagnosis of perineurioma. The classic immunohistochemical profile of extraneural perineurioma includes positivity for EMA and negativity for S100 protein, CD34, or SMA.<sup>[9]</sup>

The most important differential diagnosis of extraneural (soft-tissue) perineuriomas includes other common nerve sheath tumors like schwannomas, neurofibromas, and solitary fibrous tumor. However, most cases can be differentiated on morphology or by immunohistochemistry. The soft-tissue perineuriomas can be either encapsulated or unencapsulated. The schwannomas are encapsulated, show Antoni A and Antoni B areas, positive for S100, and stain negative for EMA. Neurofibromas are unencapsulated and show extensive S100 positivity. Solitary fibrous tumor have a patternless pattern with dense collagen bundles, hyalinized vessels, staghorn vessels, focal storiform pattern may be present, and negative for EMA.<sup>[10]</sup>

Other differential diagnoses include malignant tumors such as perineurial malignant nerve sheath tumor, malignant fibrous histiocytoma with myxoid change, and low-grade fibromyxoid

sarcoma.<sup>[11]</sup> These malignant tumors have infiltrative growth pattern, significant cytologic atypia, presence of necrosis, and high mitotic activity. Some atypical cellular features such as scattered hypercellular areas, pleomorphic or multinucleated cells, or focal infiltrative margins are also noted in few extra neural perineuriomas, but these features are considered to be a degenerative change and, therefore, to have no clinical or prognostic significance.<sup>[12]</sup> Malignant transformation has not been reported. In our case, no features of atypia or malignancy were seen.

As extraneural perineuriomas have a benign clinical course, hence surgical excision with clean margins is a treatment of choice.

## Conclusion

It is a benign, slowly growing soft-tissue mass. The definitive diagnosis is made by histologic features and immunohistochemical expression of perineurial cell markers. We have discussed this case with the main objective of making the clinicians aware of this rare entity especially when the patient is an infant where overtreatment should definitely be avoided.

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