

Atypical presentation of a cellular neurothekeoma on the nasal vestibule of a pregnant woman

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Neurothekeomas are rare benign solitary tumours of the skin, most commonly manifesting in the head and neck and upper extremities of adults within the second to third decade of life, more often appearing in females at a 2:1 ratio. The authors describe a case of an immunohistochemical diagnosis of a cellular neurothekeoma on the nasal vestibule of a pregnant woman, and discuss the curative ability of surgical excision in these patient cohorts.

Key Words: *Nasal vestibule; Neurothekeoma; Pregnant*

Neurothekeomas are uncommon benign solitary lobulated tumours of the skin, most commonly manifesting in the head, neck and upper extremities of adults in their second or third decade of life, with a proclivity for females at a ratio near 2:1 (1,2). These lesions generally manifest as indolent, asymptomatic, pigmented growths ranging in size from 0.5 cm to 3 cm without involvement of underlying subcutaneous fat or skeletal muscle (1). The broad histological appearance of these lesions has led to subcategorization into myxoid, cellular and mixed lesions. The myxoid variant is characterized by well-circumscribed stellate and spindle cells within a mucinous matrix of mucopolysaccharides, whereas the cellular form exhibits fascicular nodule aggregation into nests with a grenz zone partitioning the epidermis and adjacent tumour (2). We report a novel case of a cellular neurothekeoma (CNT) on the left nasal vestibule of a pregnant woman that exhibited atypical rapid growth after previous excision, the first case reported of local excision in an encephalic patient.

CASE PRESENTATION

A previously healthy 33-year-old woman in her second trimester of pregnancy was seen after a left vestibular lesion had reappeared after an initial excision at another institution four months previously. The lesion presented asymptotically one year previously, with steady growth and increasing discomfort after becoming pregnant. She had no previous history of bleeding, pain, ulceration, nasal obstruction or cancer. On physical examination, the tumour presented as a well-circumscribed erythematous 7 mm nodular lesion on the vestibular surface of the left ala. The pathology report from the initial excision was consistent with a neurothekeoma. Due to the benign diagnosis, a decision was made to postpone surgery until after the delivery.

One month later, the patient returned, stating the lesion had doubled in size and now was causing significant nasal obstruction (Figure 1). Sinuscopy did not reveal any synchronous lesions. Due to the tumour's rapid growth and the patient's new functional impairment, she requested surgery, which was supported by her obstetrical team.



Figure 1 Preoperative frontal view of left vestibular tumour

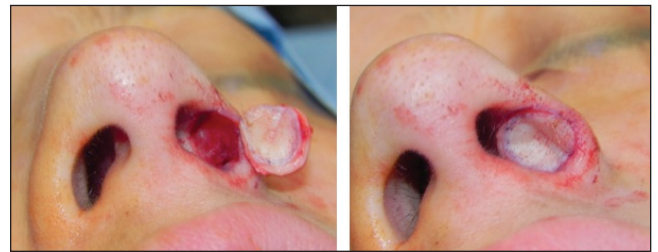


Figure 1 Excision of a nasal cellular neurothekeoma with margins (left), deficit replacement with cyma conchae composite graft (right)

Under general anesthesia, a wide local excision of the left nasal mass was performed with 2 mm to 3 mm margins. A right auricular composite graft was performed for reconstruction (Figure 2). There were no complications, her postoperative course was uneventful and she experienced immediate relief of her nasal obstruction. Pathology showed a highly mitotic polypoid mass involving the dermis, with skeletal muscle penetration, consisting of fascicles of nests forming from spindle and epithelioid cells, and an unaffected grenz zone. The morphological appearance and immunohistochemical profile of positive CD10, CD68, D240 and negative S100, indicated a diagnosis of CNT with atypical features.

CONCLUSION

CNTs are diagnostically challenging due to the lack of a distinct immunophenotype and overlapping morphology with other skin lesions including melanomas. However, the absence of the S100 protein marker is a strong indicator against melanocytic origin (3).

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Additionally, this differentiates the tumour from the myxoid variant, which presents with a septated growth pattern, prominent myxoid stroma, and S100 and GFAP protein expression (3). Furthermore, the presence of multiple associated biomarkers including MITF, CD10, CD68, and morphological characterization of lobular growths forming fascicles and a grenz zone, cumulatively consolidated a diagnosis of CNT (4).

This patient presented with atypical features due to a brisk mitotic rate, deeper penetration, infiltrative borders and cytological pleomorphism (5). Regardless, for all categories of neurothekeoma, including those with atypical features, surgical removal is curative and reoccurrence is often due to inadequate resection margins (5). The differential diagnosis for CNT include spitz nevus, melanoma, plexiform fibrous histiocytoma, plexiform neurofibroma and dermatofibroma (4). It is important to accurately classify these tumours to avoid unnecessary work-up, wider surgical margins and rigid monitoring required for more sinister diagnoses.

The aforementioned recurrence of a benign, rapidly growing, highly cellular, invasive polypoid mass on the nasal vestibule was an atypical presentation of a CNT in a pregnant woman. The present report is the first to describe the regrowth and re-excision of a CNT in a pregnant woman, fortifying the notion that these benign tumours

can present on the nasal vestibule of these particular patients. Additionally, the favourable outcome from local excision with margins reaffirms the concept that surgical removal is curative even with atypical presentation in this cohort of patients.

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