Facial neurothekeoma in a 10-year-old child

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Neurothekeoma is a rare and benign tumour that occurs most frequently on the head and neck. It is classified as a nerve sheath tumour and epidemiologically occurs in young adults. It is rare to find these tumours occurring in children. The following report describes a case of neurothekeoma in a young girl, its treatment and a review of the literature. Specific histological analysis may reveal atypical and aggressive features, which, in turn, affect resection and reconstructive options.

Key Words: *Immunohistochemistry; Nerve sheath tumour; Neurothekeoma; Reconstructive surgery*

Un neurothécome facial chez une enfant de dix ans

Le neurothécome est une tumeur bénigne rare qui se forme surtout sur la tête et le cou. Il est classé parmi les tumeurs de la gaine nerveuse. D'un point de vue épidémiologique, il se manifeste chez les jeunes adultes. On l'observe rarement chez les enfants. Le présent rapport décrit un cas de neurothécome et son traitement chez une jeune fille, et s'enrichit d'une analyse bibliographique. L'analyse histologique spécifique peut révéler des caractéristiques atypiques et agressives qui ont une incidence sur les possibilités de résection et de reconstruction.

CASE PRESENTATION

In 2004, a 10-year-old girl was referred for a workup and treatment of a 1 cm × 1 cm round lesion on the nasal dorsum. The child was otherwise healthy with no prior medical history and had had normal development. The lesion was firm and red in colour, and the edges were smooth with a clear demarcation grossly between normal skin and that of the lesion (Figure 1). It was mobile and did not cause the patient any pain. Regional lymph nodes were unremarkable. A biopsy of the lesion was performed at the initial visit and sent to pathology. A diagnosis of neurothekeoma was made, and the patient returned for definitive treatment. It was decided to perform the excision and reconstruction under general anesthesia.

METHOD

The lesion was outlined using a marking pen with a 2 mm gross margin. The defect was reconstructed with a bilobed flap (Figures 1 and 2). Deep dermal interrupted sutures with 5-0 chromic were used to approximate the flaps, followed by 6-0 Prolene (Ethicon Inc, USA) interrupted sutures to close the skin (Figure 3). The patient did very well postoperatively with negative superficial and deep surgical margins on the pathology report (Figure 4). Immunohistochemistry evaluation of the tumour was positive for vimentin, and negative for S-100, epithelial membrane antigen and NKI/C3 stains. Follow-up after two weeks and one year showed no recurrence with a good aesthetic result (Figures 5 and 6).

DISCUSSION

Neurothekeoma is a rare, benign tumour that is derived from peripheral nerve sheath cells. There are less than 300 cases reported in the literature to date. There is a greater incidence among female patients than male patients, and the tumour has been shown to occur anywhere from infancy to the age of 84 years, although young adults are the most often affected. Anatomically, the tumour occurs most frequently in the head and neck, followed by the upper extremities. Even rarer cases in lower extremities and the trunk have been reported in selected case studies. The rarest cases have been reported in the spine and on the digits of the toes (a total of five cases) (1).

Classifications of this tumour have evolved over the years. It was originally described by Harkin and Reed (2) over 30 years ago, whereby histological analysis revealed cells of endoneurium origin with a large proportion of mucoid matrix. The name of neurothekeoma ('theke' is Greek for sheath) was suggested in 1980, when Gallagher and Helwig (3) published their series and proposed the name. In 1985, the largest series was published by Pulitzer and Reed (4) and reported on 70 cases. The largest review of all cases was reported by Papadopoulos et al (1), who reviewed the location and incidence of 300 neurothekeoma cases in the pediatric and adult populations (82 known cases in children).

Histologically, neurothekeomas are classified into three groups: classical (myxoid or hypocellular), cellular and mixed types. Each has their own characteristics as seen under light

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Figure 1) Neurothekeoma excision margins and planned bilobed flap

Figure 2) Lifting bilobed flap

Figure 3) Closed bilobed flap

microscopy and on immunohistochemistry patterns. Busam et al (5) offer an accurate description of the morphological features associated with neurothekeoma. It consists of cellular fascicles

Figure 4) Nests of neurothekeoma cells are often seen in close relationship with small nerves (arrow). Note the myxoid matrix (bluish tint) characteristic of neurothekeoma cells and the absence of cytological atypia and mitotic activity, features of benignity (hematoxylin and eosin stain)

Figure 5) Close-up of flap at two-week follow-up visit

Figure 6) Close-up at one-year follow-up visit

and circular swirls of spindle and epithelioid cells, and a background composed of myxoid or sclerosing substance. The tumour infiltrates in a plexiform fashion. The different histological subtypes are categorized in relation to their cellularity, mucin content and growth pattern. The hypocellular myxoid type usually shows encapsulation and sharp demarcation. Immunohistochemistry stains positive for S-100, collagen

type IV and frequently epithelial membrane antigen, although exceptions to this rule have been reported (6-8). The cellular type contains scarce mucin and nodular or infiltrative growth patterns. Immunohistochemistry also shows positive reaction for neuron-specific enolase, Leu-7 and smooth muscle-specific actin (6). However, the cellular type also consists of lobulated dermal tumour composed of spindle and epithelioid cells, with a fascicular arrangement, but usually is NK1/C3 positive (5). This variant almost always stains negative for S-100 (9-11). The mixed type of neurothekeoma shows variable mucin content and cellularity, and unreliable immunohistochemical markers. A recent article by Rudolph and Schubert (12) underlines the importance of using immunophenotyping rather than light microscopy features alone to classify a neurothekeoma in the proper category. Similar studies abided by a similar philosophy (7,8).

Neurothekeomas are classically a benign tumour; however, atypical histological features can make them behave aggressively and extend into fat or skeletal muscle, and one case revealed vascular invasion (5). However, surgical resection is almost invariably curative except in a few cases in which the tumour showed recurrence (5). This, however, was due to incomplete margins of resection. There are no reports in the literature of metastases. Busam et al (5) and Bhatia et al (13)

summarize the characteristics of atypical findings as large size (to 6 cm), extension into muscle or fat, vascular invasion, infiltrative borders, high mitotic index and distinct pleomorphism. There are no guidelines for standard resection margins, but it would seem that a few millimeters from grossly detectable margins is usually sufficient. However, due to the occurrence of atypical neurothekeomas, a punch biopsy to confirm this atypia may be prudent before embarking on reconstruction. If atypical and aggressive features are found within the histological analysis, waiting for permanent margins may be advantageous. With regard to rotation or transpositional flaps of the face, recurrence can have significant repercussions in tissue coverage and planning. In our case, the histology revealed a nonaggressive, typical neurothekeoma, and therefore, gross margins of 2 mm were sufficient. There has been no recurrence, and the reconstruction was performed immediately after resection.

CONCLUSIONS

Neurothekeomas are infrequent lesions, but when they occur, specific histological analysis and immunophenotyping may be needed if any signs of aggressive behaviour are associated with the lesion. Atypical, aggressive tumours may need a two-step approach to wait for permanent margins before embarking on reconstructive efforts.

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