CASE REPORT

Infiltrating giant cellular blue naevus

A L Bittencourt, D A Monteiro, O J De Pretto

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Introduction: Cellular blue naevi (CBN) measure 1–2 cm in diameter and affect the dermis, occasionally extending into the subcutaneous fat. The case of a 14-year-old boy with a giant CBN (GCBN) involving the right half of the face, the jugal mucosa and the lower eyelid with a tumour that had infiltrated the bone and the maxillary and ethmoidal sinuses is reported.
Methods: Biopsies were taken from the skin, jugal mucosa and maxillary sinus. The following markers were used in the immunohistochemical evaluation: CD34, CD56, HMB-45, anti-S100, A-103, Melan A and MIB-1.
Results: The biopsy specimens showed a biphasic pattern affecting the lower dermis, subcutaneous fat, skeletal muscle, bone, jugal mucosa and maxillary sinus, but there was no histological evidence of malignancy. The tumour cells were CD34−, CD56−, HMB45+, anti-S100+ and A-103+. Melan A was focally expressed. No positive MIB-1 cells were identified.
Discussion: The present case shows that GCBN may infiltrate deeply, with no evidence of malignancy.

Cellular blue naevi (CBN) generally measure 1–2 cm in diameter and are present at birth in one third of cases. They usually include the reticular dermis, occasionally penetrating into the subcutaneous fat. Histologically, they are characterised by increased cellularity and a dual cell population of pigmented dendritic cells and fusiform cells arranged in various configurations and with varying degrees of melaninisation.[1] More extensive CBNs have been referred to as large plaque or giant CBNs (GCBNs).[2–5] The GCBN can be locally aggressive and may infiltrate subjacent structures, although it results in no malignant transformation.[3–10] Here, we present a case of facial GCBN that infiltrated deep structures.

CASE REPORT

A 14-year-old white boy was born with a blue macula affecting the right inferior eyelid and the subpalpebral region. According to his mother, the lesion began to grow in size when he was 4 years old and began to infiltrate 7 years prior to presentation. No mongolian spot was detected at birth. Other than his facial appearance, the child had no complaints.

Dermatological examination showed a wide, blue-coloured lesion, measuring 10 cm×11 cm at its maximum diameter, occupying the right half of the face. The lesion was infiltrated and presented some isolated nodules and several small, dark-blue papules measuring 0.2–0.3 cm in diameter. A hard tumour, measuring 4.5 cm×3.5 cm, was present in the lower right eyelid. The upper eyelid and the supra-palpebral area also showed a blue discolouration. Oral examination showed an extensive, infiltrated blue lesion in the right jugal mucosa. The child has severe facial asymmetry (fig 1). No sign of any other blue naevus was seen on his body. No superficial lymphadenopathy was detected. A bluish discolouration was observed in the bulbar and tarsal conjunctivae. No other abnormalities were seen in the ophthalmological examination. Visual acuity and tonometry were normal. Chest *x* ray was normal. Magnetic resonance imaging and tomography showed infiltration of the retrobulbar and periorbital soft tissues and the orbital bone, with a widening of the orbital fissure. The tumour filled the right maxillary sinus, projecting into the nasal cavity. The ethmoidal sinus on the same side had also been infiltrated (fig 2). The optic nerve and the extrinsic ocular muscles were not infiltrated. Owing to the degree of infiltration of the tumour, this case was considered inoperable. After 8 months of follow-up, the child had no other complaints.

Pathology

Biopsy performed from the tumour of the lower eyelid showed melanin-laden dendritic melanocytes and amelanotic or

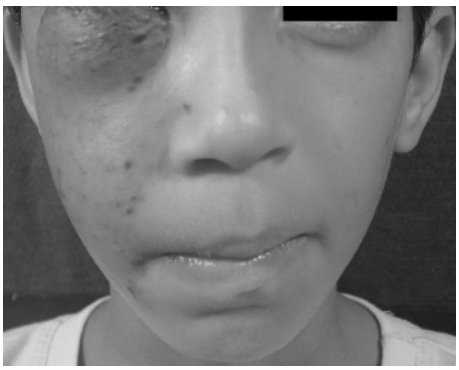

Figure 1 Facial asymmetry caused by an infiltrative giant blue naevus. Presence of a tumour in the lower eyelid.

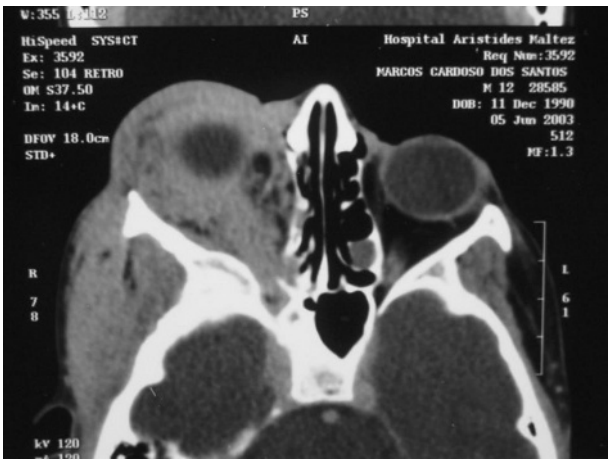

Figure 2 Tomography (section of 5 mm at the level of the orbital region). Note infiltration present in the retrobulbar and periorbital soft tissues, in the orbital bones and in the right maxillary and ethmoidal sinuses.

Abbreviations: CBN, cellular blue naevi; GCBN, giant cellular blue naevi

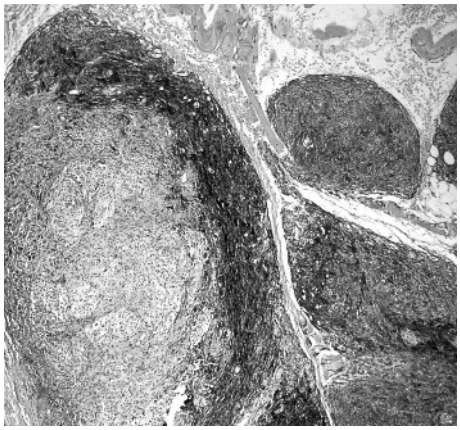

Figure 3 Tumour of the maxillary sinus showing area of increased cellularity, but without cellular pleomorphism. Haematoxylin and eosin, ×200.

paucimelanotic fusiform cells with eosinophilic cytoplasm arranged in varying patterns (fig 3). The tumour affected the lower dermis, the subcutaneous fat and the subjacent skeletal muscle. Highly cellular nests of amelanotic round cells with vacuolated cytoplasm, and vesicular nuclei with small nucleoli were seen (fig 4). No mitosis, necrosis or nuclear pleomorphism was seen. Another biopsy was performed through the jugal mucosa including bone and the tumour of the maxillary sinus. Histologically, appearance of the jugal mucosa was similar to that of the biopsy described earlier. The bone marrow was infiltrated with heavily pigmented melanocytes (fig 5) that after blanching with potassium permanganate showed cells with round or oval bland nuclei and vacuolated cytoplasm. The tumour showed compactly distributed, non-pigmented and non-pleomorphic fusiform cells, the nuclei of which had no prominent nucleoli (fig 6). No necrosis or mitosis was observed. Immunohistochemical analysis of the tumour in the non-pigmented areas, using the immunophosphatase technique (streptavidin–biotin system), showed that the tumour cells were MIB-1-and CD34-negative but were positive with HMB45, anti-S-100 protein and A-103. Melan A was weakly and focally expressed in the cells of blue naevus. In 10 high-power fields (40× objective) no positive MIB-1 cells were identified in the three biopsies (fig 7).

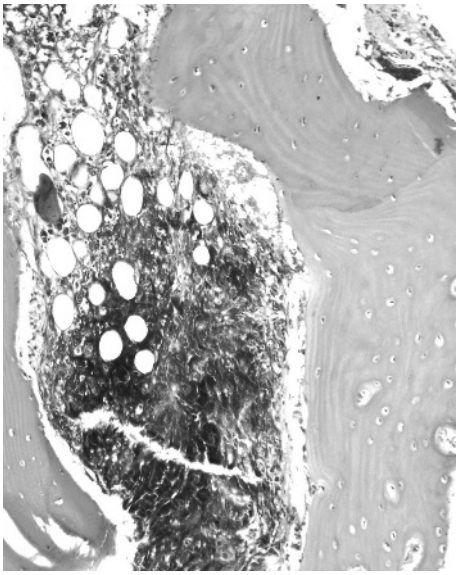

Figure 5 Bone marrow infiltrated by heavily pigmented melanocytes. Haematoxylin and eosin, ×50.

COMMENTS

This report is of a patient who had an infiltrating GCBN with no histological characteristics of malignancy including increased mitotic rate, necrosis, nuclear atypia, pleomorphism and prominent nucleoli.[11] Moreover, the mitotic index calculated using MIB-1 was negative. Smith *et al*[12] described CD34-positive CBNs without malignancy that had infiltrated deeply into the subcutaneous fat. They suggested that these naevi arise from more primitive, neurocristically derived cells; however, in the present case, the tumour was CD34-negative. One interesting aspect observed in our case was the absence of infiltration in the internal structures of the eyeball and of the optic nerve.

It is known that CBN may in rare cases lead to malignant blue naevi.[13] There is one report in the literature of an extensive periorbital CBN that presented evidence of low-grade melanoma in a few fields of some biopsy specimens.[14] Considering the extension of the lesion in this case, it is difficult to exclude the possibility of malignant transformation in focal areas. However, the literature has reports on other cases of GCBN

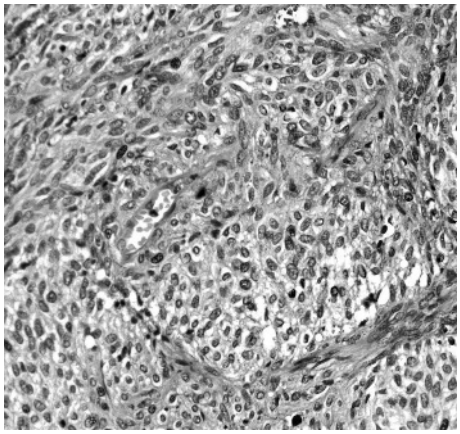

Figure 4 Greater magnification of fig 3. Round cells compactly disposed. Haematoxylin and eosin, ×200.

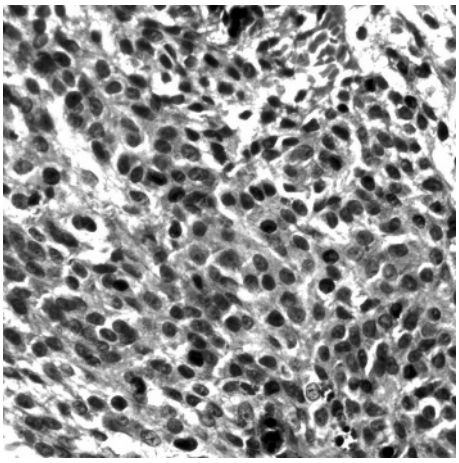

Figure 6 Tumour of the maxillary sinus showing increased cellularity but without cellular pleomorphism. Haematoxylin and eosin, ×320.

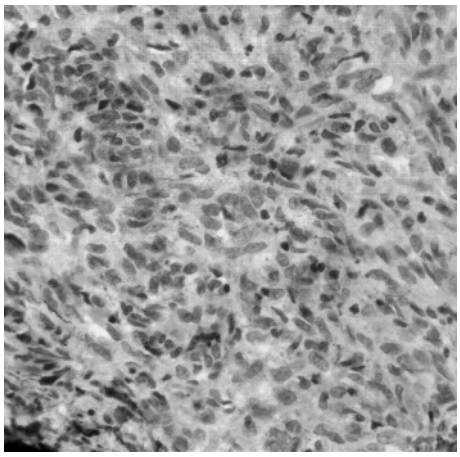

Figure 7 Cells of the tumour of the maxillary sinus. No nuclei stained with MIB-1, ×320.

with infiltrating capacity but without malignancy. When present in the scalp, GCBN may infiltrate the soft tissue, bone and dura mater, and may even form intracranial masses.[4–6 9] When GCBN is located in the anterior thorax, infiltration of the breasts may occur.[3 7] Therefore, it is important to follow up cases of GCBN carefully because it may infiltrate deeply and may even develop into melanoma.

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Written consent has been obtained from the mother of the patient to publish this data.

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