Giant neurocristic hamartoma of scalp: a case report

Arunkumar Baskara, Nancy Sapanara and Lisa Medvetz

Department of Surgery, Mercy Catholic Medical, 51–7 Revere Road, Drexel Hill, PA 19026, USA

Corresponding address: Dr Arunkumar Baskara, Department of Surgery, Mercy Catholic Medical, 51–7 Revere Road, Drexel Hill, PA 19026, USA.
Email: abaskara24@yahoo.co.uk

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Abstract

Neurocristic hamartoma results from the aberrant development of cells derived from the neural crest. It is a type of dermal melanocytosis. These tumours may contain neuro-sustentacular and fibrogenic components in addition to a dermal melanocytic component. The elements within these tumours reflect the spectrum of differentiation that results from migration of neural crest-derived cells. Identification of these tumours could be important if malignant transformation results in the development of tumours with a distinctive biologic behaviour.

Keywords

Neurocristic hamartoma; scalp tumour.

Case report

The patient was a 36-year-old man who had recently migrated to the United States from Kenya. He presented to our outpatient office with a large lesion in the right parieto-occipital area of the scalp (Fig. 1). The lesion started as a small lump in the scalp and gradually grew in size over a year. It was painless and was producing an intermittent serous discharge, which was worse at night. He had no significant past medical or surgical history, and no history of exposure to tuberculous patients. He was not on any medications. At the time of his presentation, the lesion was 11 cm in diameter, with patchy areas of superficial skin necrosis. The lesion was dry. Routine laboratory investigations were normal.

A computed tomography (CT) scan of the head showed a scalp tumour with no definite evidence of osseous erosion. An magnetic resonance imaging (MRI) scan of the head revealed possible erosion of the outer table of the skull adjacent to the lesion but no intracranial involvement. A metastatic workup, including CT of the neck and chest, and a positron emission tomography (PET) scan were done. These revealed non-significant right-sided submandibular and jugular lymph nodes but no other abnormalities. An incision biopsy of the lesion was carried out electively. The histopathology report showed a malignant spindle cell soft tissue tumour. After extensive discussion with the patient, a wide local excision of the lesion was carried out including a 2 cm margin of skin around the lesion and excision of all deep tissue as far down as the scalp periosteum. The scalp defect (Fig. 2) was closed temporarily with a superficial split skin graft and

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a vacuum dressing was applied. The patient was discharged home on the fourth post-operative day with the vacuum dressing in place.

Pathological examination of the excised specimen revealed a widely infiltrative spindle cell lesion with focal fibrosis, nuclear palisading, and pigmentation. No necrosis or mitotic activity was present. There were elements of a blue nevus-like histology, schwannian nerve-sheath like changes, abnormal hair formation, and dendritic pigmented cells. Nuclear staining by immunohistochemistry was positive for microphthalmia-associated transcription factor (MiTF) (Fig. 3) and immunostains for S-100 and Melan-A were also positive. The immunohistochemistry results were suggestive of a neurocristic hamartoma. A flap closure of the scalp defect is planned and will be carried out in the plastic surgery department.

Discussion

Neurocristic hamartoma (NCH) results from the aberrant development of neuromesenchyme. Thus, the elements within these tumours reflect the spectrum of differentiation that results from
migration of neural crest-derived cells. This was first described in 1982 by Tuthill et al.[1] These cells show fibrogenic, melanogenic and neurosustentacular differentiation and result in the formation of the local mesenchyme in the cephalic regions[2]. These lesions have a tendency to transform into malignant melanoma over an unpredictable period of time[3].

NCH is a type of dermal melanocytosis. Dermal melanocytosis includes a variety of pigmented lesions that are formed from the aberrant development of neural crest-derived melanocytes as they migrate to the dermis during embryogenesis[4].

NCH is composed of melanocytes that are confined to the dermis and sometimes the subcutaneous tissue with a neural crest-derived Schwann cell component[5]. NCH has a predilection for the scalp, perhaps because it is associated with focal alopecia. It is generally not found on the trunk. The features of NCH overlap with blue nevus both clinically and histopathologically. Clinically, NCH are large lesions from 3 to 10 cm in diameter compared with blue nevi, which are typically much smaller (1–3 cm).

Histologically, NCH lesions are composed of dermal melanocytes and neuroidal structures with schwannian differentiation. These lesions stain with CD34, whereas blue nevi usually do not. The number of hair follicles in NCH lesions are decreased, as mentioned above, but not in the blue nevus. Immunostaining in our patient confirmed a melanocytic-nevic origin and the lesion was floridly positive for S100, MiTF, Melan A, tyrosinase, CD34 and HMB45.

Melanomas have been reported to develop at any time between 1 and 6 years from the initial diagnosis of NCH[6]. In congenital NCH, melanomas have been identified 15 to 60 years after birth[6]. Long-term follow-up with cancer surveillance is recommended in patients with NCH. Complete resection of the lesion prevents the development of melanoma.

NCH of the scalp is a rare tumour. Early diagnosis if the exact nature of the tumour is crucial for effective management. Although our patient presented with a giant NCH of the scalp, the pathology results showed no evidence of melanoma in the lesion and there was no evidence of metastatic tumour on thorough investigation.

Teaching point

Because NCH may resemble a blue nevus both clinically and histopathologically, it should always be considered in the differential diagnosis, despite its rarity. Early diagnosis and treatment is mandatory because of the possibility of malignant melanoma developing in these lesions. Surgical excision should be considered for reasons of cosmesis or for lesions thought to be at increased risk of malignancy and/or too difficult to follow clinically as an outpatient.

References


