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Langerhans cell histiocytosis (eosinophilic granuloma) of the skull mimicking nummular headache. Report of two cases

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Abstract

Background: Nummular headache is a rare, recently described topographic headache defined by the circumscribed coin-shaped area of pain. It is classified as a primary headache. There is debate about whether it is due to a peripheral or central disturbance, and its relationship to migraine.

Case reports: We report two patients with presumed nummular headache secondary to Langerhans cell histiocytosis, both with resolution of their headaches after surgical resection.

Conclusion: Imaging in patients with clinical features of nummular headache is recommended, as this and other cases highlight that it may be symptomatic. There are no distinguishing clinical features to separate nummular headache from secondary mimics, and treatment of the underlying cause may be curative.

Keywords

Nummular headache, coin-shaped cephalgia, Langerhans cell histiocytosis, eosinophilic granuloma, topographic headache

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Background

Nummular headache (from the Latin *Nummus*: coin) is a rare headache, with a hospital incidence of 6.4 cases/100,000 (1), accounting for 6% of unilateral headaches (2). It was described by Pareja et al. in 2002 (3) and has been incorporated in the International Headache Classification (ICHD) since 2004 (4). It is characterised by a circumcised scalp pain in the shape of a coin or an ellipse of 1–6 cm in diameter. The pain is most often localised to the parietal region and may co-exist with sensitivity of the skin in the area of the pain. The pain is periodic, of moderate intensity and usually presents with a duration of more than three months. There may be periods of remission, but exacerbations of variable durations are well described (5). There are no randomised treatment trials, but gabapentin is the most common of the many reported treatments (5).

Nummular headache is classified as a primary headache disorder, defined by topography along with primary stabbing headache and epicrania fugax as epicranial headaches (head pain over the scalp). Absence of a secondary cause is a diagnostic pre-requisite. There is debate about whether it has a central or peripheral mechanism (6). The minority of patients have co-existing migraine, which runs a separate course (1).

Although the absence of a specific structural lesion is a criteria for the ICHD diagnosis, there are a few reports of headaches with the clinical features of nummular headache but with structural cases (7–10) (see Table 1).

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Table 1. Current and previously reported cases of secondary headaches with clinical features of nummular headache.

Lead author	Year	Age (years)/sex	Aetiology	Treatment	Comments
Silva-Rosas[a]	2017	35, M	Langerhans cell histiocytosis localised to a single area of the cranium	Resection	Resolution post resection
		12, M	Langerhans cell histiocytosis localised to a single area of the cranium	Resection	Resolution post resection
Chui (10)	2013	52, F	Pituitary oncocytoma	Resection	Resolution post resection
Álvaro (7)	2009	67, M	Benign lesion protruding from the skull, with preserved bone	Local infiltration with lidocaine	Medication response poor
		72, F	Pituitary adenoma	Eighteen years after trans-sphenoidal approach	Medication response poor
Guillem (8)	2009	36, F	Arachnoid cyst	Surgery declined	Medication response poor
		52, F	Arachnoid cyst	Surgery declined	Partial response to pregabalin
Guillem (9)	2007	60, F	Subtentorial meningioma	Resection	Resolution post resection

M, male; F, female.
[a]Present cases.

Clinical cases

Case 1

A 35-year-old Amerindian male patient presented with a four-month history of pain restricted to a 5 cm diameter zone in the right parietal region. There was no past medical, migraine or headache history. The pain intensity was 5/10 (11) and described as a continuous sensation of ice. He had a partial response to gabapentin at a dose of 900 mg per day. He fulfilled the clinical criteria of nummular headache; however, a computed tomography (CT) scan and magnetic resonance imaging (MRI) (Figure 1a) showed a lesion underlying the symptomatic area. Skeletal studies, immunoglobulin level and paraprotein results were normal. The patient had a craniectomy with a margin of healthy bone and a cranioplasty with acrylic. Histopathology showed eosinophilic granuloma (histiocytosis) with complete resection. Postoperatively, the patient was immediately free of pain and was fully recuperated two days after surgery. Over the following five years he has remained well without any symptoms.

Case 2

A 12-year-old Hispanic male patient with no relevant medical or headache history presented with a three-month history of circumscribed pain in a 3 cm oval in the left lateral frontal region. The intensity of the pain was 3/10, but contact with the sensitive area, for example when 'heading' a ball playing soccer, intensified the pain to 9/10. Examination of the area was normal, with no swelling and no skin lesions, apart from allodynia in the affected area. The patient fulfilled the clinical features of nummular headache. However, skull radiography, CT scan and MRI showed a circumscribed lesion underlying the symptomatic area (Figure 1b). Skeletal survey, immunoglobulins and protein immunoelectrophoresis were normal. A craniectomy was performed leaving a healthy bone margin. The biopsy showed an eosinophilic granuloma (focal histiocytosis) with complete resection. Pain relief was immediate and the patient is well and pain free at six months' follow-up.

Discussion

These patients are the first described cases of presumed nummular headache secondary to Langerhans cell histiocytosis (eosinophilic granuloma). They otherwise meet all the clinical features of nummular headache, and did not fulfil the characteristics of paroxysmal or continuous hemicrania or other lateralised headache syndromes. These patients did not try indomethacin or other medications, apart from gabapentin, as they proceeded to curative surgery when the underlying lesion was found.

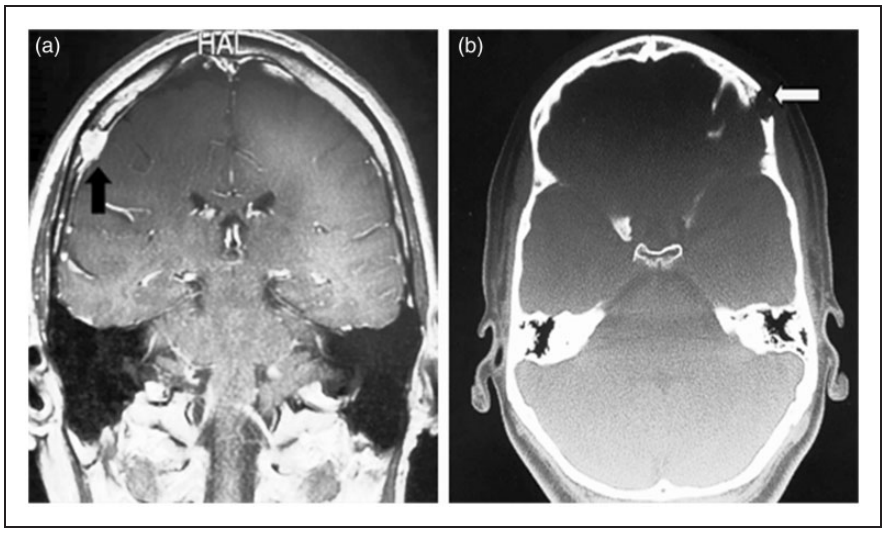

Figure 1. (a: Case 1) Post-gadolinium DTPA T-weighted coronal magnetic resonance imaging (MRI) demonstrating an eosinophilic granuloma (black arrow). (b: Case 2) Bone window of an axial computed tomography (CT) scan showing a lytic lesion–eosinophilic granuloma (white arrow).

There are a few previously reported secondary causes for presumed nummular headache, such as arachnoid cysts and subtentorial meningioma. Both of the patients described here had Langerhans cell histiocytosis localised to a single area of the cranium, with no evidence of dissemination at presentation or follow-up of five years and six months respectively. Surgery was curative as resection with a clear margin led to complete and definitive pain relief, implying a peripheral mechanism for their pain. This cannot necessarily be extrapolated to other patients, but is in keeping with the view that the clinical features of nummular headache arise as a local pain disorder originating in terminal branches of a sensory nerve and inducing peripheral sensitisation in one or several primary sensory neurons (6).

There were no clinical features that differentiated these secondary head pains from cases of primary nummular headache. These cases suggest that neuroimaging should be performed in all patients with clinical features of nummular headache to exclude a structural cause for the headache.

Clinical implications

- Two patients with presumed nummular headache secondary to Langerhans cell histiocytosis had complete resolution of their headaches after surgical resection.
- Primary nummular headaches and presumed nummular headaches with a secondary cause are clinically indistinguishable.
- Neuroimaging should be performed in all patients with presumed nummular headache.

Declaration of conflicting interests

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