

Meningeal calcification in basal cell nevus syndrome

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DESCRIPTION

A 20-year-old woman presented with history of headache for 1 year. The headaches were mild to moderate in intensity, bilateral and non-throbbing, with no symptoms suggestive of raised intracranial pressure. The patient had hyperpigmented skin lesions for 2 years and she had undergone enucleation of odontogenic keratocyst of anterior mandible 1 year earlier (figure 1A).

On examination multiple hyperpigmented papules and few hyperpigmented plaques were seen over her face, neck, trunk, bilateral upper limbs, lower limbs and axilla. Some of them had rolled out border and central clearing. The patient's palms had multiple pits (figure 1B). Dermoscopy revealed blue grey globules, phylloid pattern, few spoke wheel areas and brown halos. An X-ray of the lumbo sacral spine showed bifid fifth Lumbar (L5) spinous process and scoliosis. The cerebral CT showed calcification of the falx cerebri (figure 1C) and tentorium cerebelli (figure 1D). The patient was diagnosed as having basal cell nevus syndrome (BCNS).^{1 2} None of her family members had similar illness.

BCNS is a rare autosomal dominant disorder with complete penetrance and variable expressivity. Kimonis *et al*¹ have devised criteria for the diagnosis of BCNS. Cerebral calcification is a common finding in the cerebral CT scan of the patients from Indian subcontinent because of the high prevalence of tuberculosis and neurocysticercosis. However, it is prudent for a neurologist to examine the skin and the skeletal system of young patients with meningeal calcification. Indeed it is important to identify this condition early as these patients are prone

Learning points

- ▶ Basal cell nevus syndrome is a rare autosomal dominant disorder with skin, skeletal, dental and central nervous system involvement.
- ▶ Cerebral CT scan shows calcification of falx cerebri and tentorium cerebelli.
- ▶ Patients are prone to develop multiple early basal cell carcinomas.

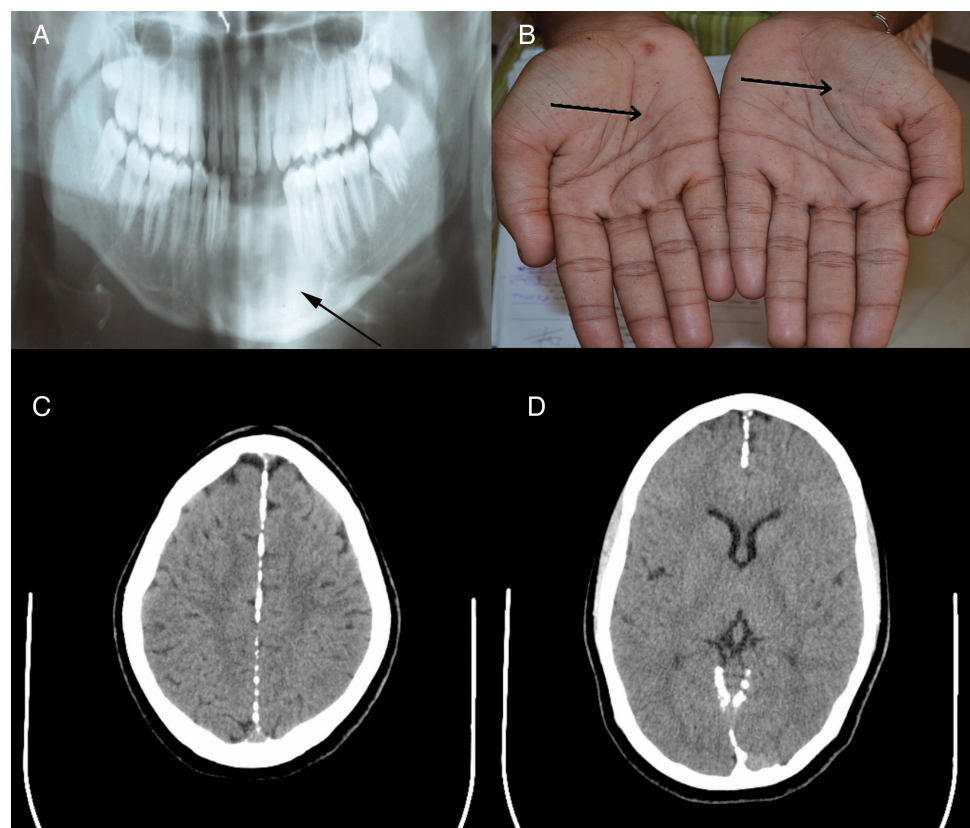


Figure 1 (A) X-ray mandible showing odontogenic keratocyst, (B) hands of the patient showing multiple pits, (C) cerebral CT scan showing falx cerebri calcification and (D) cerebral CT scan showing tentorium cerebelli calcification.



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to develop basal cell carcinomas and other tumours. Important aspects in the management of BCNS are frequent examination, counselling about sun protection, genetic counselling and early treatment of small tumours by surgical or non-surgical methods.

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