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CASE REPORT

CNS germinoma with extensive calcification: An unusual histologic finding

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Abstract

Introduction: Intratumoral calcification is a feature that is more often observed in pineal parenchymal tumour than germinoma. We describe a 13-year-old male with pineal region germinoma demonstrating extensive intratumoral calcification. *Case report:* He presented with worsening headache that was associated with fatigue, nausea and vomiting. Radiologic examination revealed a multilobular mass in the pineal region with internal calcifications. Biopsy showed a pure germinoma with unusually extensive calcification. *Discussion:* Although a diagnosis may be suggested with a careful evaluation of imaging, there is no pathognomonic pattern. Thus, histologic verification is necessary for most pineal region masses.

Keywords: Germinoma, germ cell tumour, calcification, CNS, paediatric

INTRODUCTION

Germinoma is the most common tumour of the pineal region. The main differential diagnosis is pineal parenchymal tumours. Calcification is a feature that is more often observed in pineal parenchymal tumours while in germinomas this is thought to be rare to minimal and appears as a speckled, ring-like, or stippled pattern in imaging studies when present.[1,2] So far, there are no reported cases of pure germinoma of the pineal region with diffuse calcification. Herein we describe a 13-year-old male with pineal region germinoma demonstrating extensive intratumoral calcification.

CASE REPORT

This is a 13-year-old male with a 6-week history of worsening headaches associated with fatigue, nausea and vomiting. His cerebrospinal fluid showed elevated beta-human chorionic gonadotropin (β-hCG) and normal α-fetoprotein. Radiologic examination revealed a multilobular mass in the pineal region measuring 20 x 18 x 11 mm with internal calcifications. Biopsy showed pure germinoma with large atypical cells with round nuclei, prominent nucleoli and variable clear to eosinophilic cytoplasm. Heavy intratumoral calcification (Fig. 1), and patchy-focal lymphocytic and neutrophilic infiltrates were seen. The tumour cells expressed Oct4, CD117 (cKit), PLAP and Sall4. The Ki-67 proliferation index was nearly 100%. β-hCG showed scattered staining. CD30, α-fetoprotein and Glypican-3 were negative (Fig. 2).

DISCUSSION

Pineal gland is shaped like a pine cone, located midline and attached to the posterior end of roof of third ventricle. It secretes melatonin hormone, which regulates circadian rhythm. Physiological calcification can occur in the pineal gland and this is associated with aging.[3] Calcification is significantly higher in individual with Alzheimer disease and was found to be associated with migraine.[4,5]

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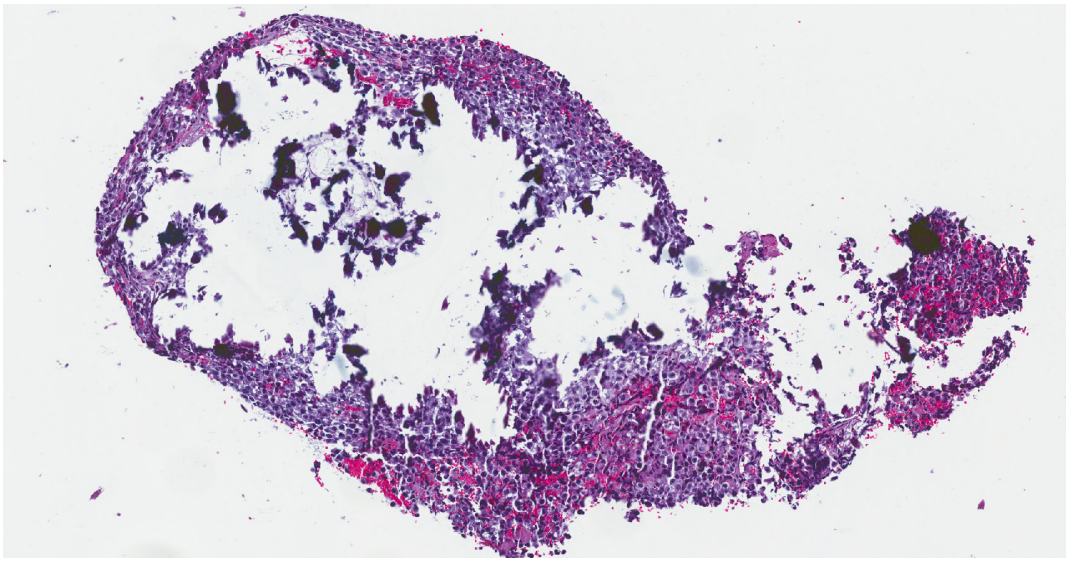

FIG. 1: The tumour demonstrated extensive intratumoral calcification (H&E, ×4).

Pineal lesions account for 1% of intracranial tumours in adults and 3–8% in children. Germ cell tumours are the most common neoplasm in the pineal region. Other tumours include pineocytoma, pineal parenchymal tumour (PPT) of intermediate differentiation and pineoblastoma from pineal parenchyma, and dermoid cyst, lipoma, meningioma and astrocytoma from nearby tissues. The majority of germ cell tumours are germinomas, followed by teratomas.[6]

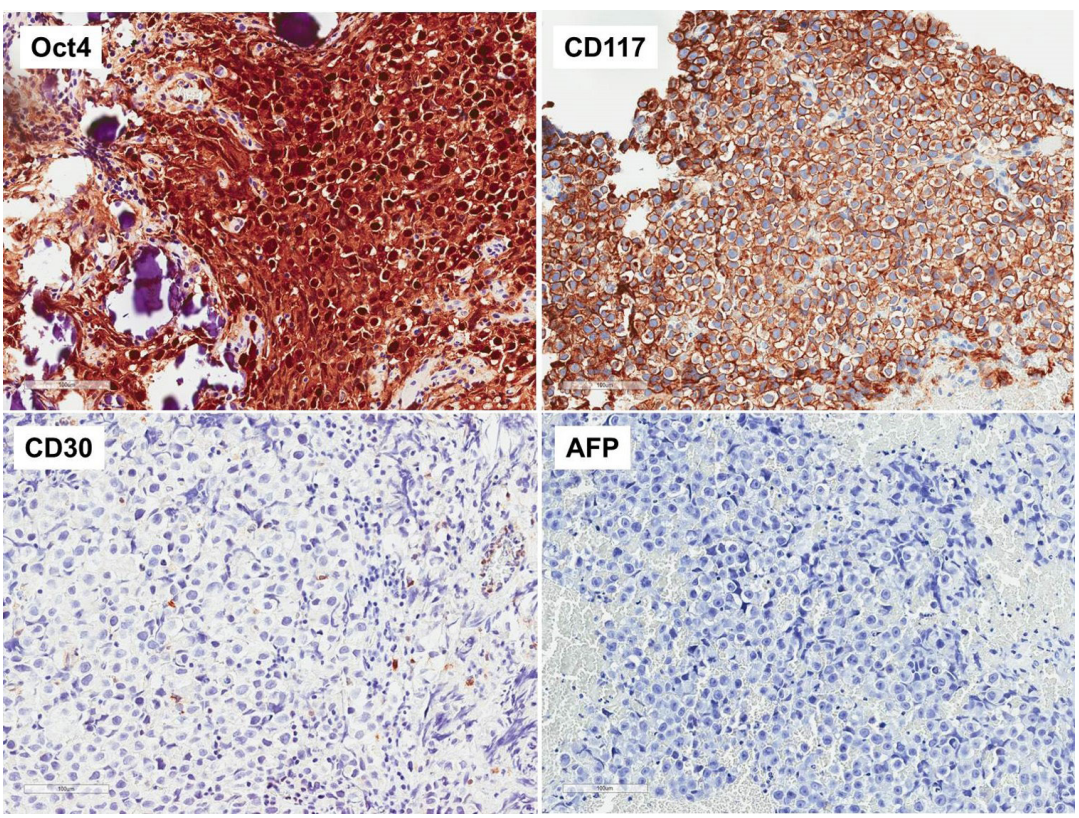

FIG. 2: The tumour cells expressed Oct4 and CD117 (cKit) (scale bar 100 μm), and are negative toward CD30 and AFP (scale bar 100 μm).

Germinoma accounts for 3-5% of paediatric intracranial tumours, 0.4-1% of adult intracranial tumours, and 50-70% of all pineal neoplasms.[3] Most patients are less than 20 years old with a male preponderance.[6] Histologically, it is characterised by variable proportions of cellular sheets or lobules of uniform germinoma cells with large round nuclei, prominent nucleoli, and clear cytoplasm interspersed by septal bands of connective tissue rich in capillaries, lymphocytes, and at times, granulomas.

Pineal calcification is a feature that is more often observed in pineoblastoma and PPT, and calcifications that tend to appear as "exploded" at the periphery, by imaging. In contrast, intratumoral calcification in germinoma is rare and if present, it adopts a "speckled" pattern. Other pineal region tumours that may have calcifications are teratomas and meningiomas. The patterns of calcification, either as "engulfed" (tumour expanding around a central area of calcification) or "exploded" (calcification that dispersed at the periphery of tumour), are distinguishing features of germinoma and pineoblastoma/PPT.

One study reported that 82% of pineal parenchymal tumours had an exploded pattern of calcification, while 78% of germinomas had an engulfed pattern of calcification.[7] Another study of 20 cases of pineal region tumours reported only one pineoblastoma that had diffuse calcification, and imaging cannot clearly distinguish between pineoblastoma and germinoma.[8] The extensive intratumoral calcification within apparently pure germinoma in this patient is unusual, and should be taken into consideration, and not mistaken as a pineoblastoma/PPT. Although a diagnosis may be suggested with a careful evaluation of imaging, there is no pathognomonic pattern. Thus, histologic verification is necessary for most pineal region masses.[9]

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