Intracerebral metaplastic meningioma with prominent ossification and extensive calcification

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Abstract

We present a patient (male 26 years) with a short history of recurrent seizures induced by a largely intracerebrally located frontal lobe meningioma. The tumor displayed a hereto-fore unpublished combination of extensive metaplastic bone formation and prominent non-psammomatosus calcifications with focal chicken-wire pattern.

Introduction

Meningiomas are relatively common tumors derived from arachnoidal cells and most frequently occur in association with intra-cranial meninges. They make up about 30 per cent of primary brain and central nervous system tumors.1 Whilst psammomatous meningiomas may be associated with metaplastic bone formation, metaplastic meningiomas with significant ossification are distinctly uncommon. In this study, we present a metaplastic meningioma which in addition to widespread ossification also exhibited extensive calcifications which focally displayed a distinctive pattern of non-psammomatosus (chicken-wire) calcification. This combination of histological features has, to the best of our knowledge, not been reported previously.

Case Report

The patient is a previously healthy 26-year-old man who developed recurrent generalized tonic-clonic seizures over a period of 3 months. A magnetic resonance imaging (MRI) of the brain showed a 4 cm large, rounded, heavily calcified tumor predominantly located in the brain parenchyma (right frontal region), causing underlying edema and left sided midline shift.

The tumor was attached to the dura by a very thin tail (Figure 1). The tumor was excised.

Discussion

Whilst all meningiomas show characteristic ultrastructural features of well-formed desmo-
expressed in the more common meningothe- 
lial meningiomas. It is also not clear if the 
ossification process in psammomatous meningi-
gomas and metaplastic meningiomas are sig-
ificantly different, although they are classi-
fied as distinct histologic subtypes. To the best 
of our knowledge, the vast majority of reported 
cases of extensively ossified meningiomas 
have contained psammoma bodies.3-6 However, 
one case of a meningioma (in the lacrimal fossa) 
which showed osseous metaplasia without 
significant non-osseous calcifications is 
on record.7 In our case, the extensive calcium 
deposits formed crystalline structures rather 
than the characteristic concentric laminations 
of psammoma bodies and the basophilic calcifi-
cations transitioned seamlessly into woven 
bone in many areas. An interesting finding in 
the tumor presented herein was the chicken-
wire-type calcifications. This pattern of calcifi-
cation is very similar to what is commonly 
encountered in chondroblastomas (CB) and 
has not previously been reported in a meta-
plastic meningioma. Chondroblastomas are 
rare primary bone tumors (1% of all primary 
bone neoplasms) which most commonly occur 
in the epiphysis of long bones with only 1% 
seen in the skull (Kobayashi Y 2001). 
Especially on small biopsy specimens with a 
paucity of lesional cells, the rare examples of 
the latter category may pose differential diag-
nostic difficulties to a MM where this type of 
calcification pattern is encountered. However, 
paying close attention to the morphological 
features of CB should help to resolve this. CB 
has a characteristic matrix (pink cartilage) 
and the lesional cells are rounded-epithelioid 
with round to ovoid grooved nuclei and the 
presence of multinucleated giant cells in CB 
are in contrast to the whorls and spindly mor-
phology of most meningotheelial cells which 
also frequently display nuclear inclusions. Of 
note is that immunohistochemically, in addi-
tion to expressing S-100 protein, chondroblas-
tomas have been shown to express EMA.8 
Osteoblastomas may arise both in the perioste-
um of the calvarium (calvarial periosteal 
osteoblastomas9,10 and in an extra-osseous and 
intracranial locations with similar radiologic 
features as a meningioma.11 This is especially 
important since in one case located in the cal-
varium, EMA expression was detected.9 
Another differential diagnosis that should be 
contemplated is the calcifying pseudotumor of 
the neural axis. This rare lesion presents as a 
granular mass located in the meninges and is 
characterized by amorphous calcifying materi-

Figure 2. A heterogeneous tumor with areas of calcification and bone formation. (A) 
Tumor seen at low power showing cellular areas with areas of calcification and ossi-
faction. (B) The cellular areas are composed of plump, elongated cells with meningothe-
elial features. (C) Prominent lymphoplasmacytic infiltrate is present within the cellular 
areas. (D) The highly vascularized tumour also contain dysplastic blood vessels. (E) The areas 
of calcification show crystalline deposits of calcium merge seamlessly with woven bone. 
No osteoblastic rimming is seen. (F) An interlacing chicken-wire-like calcification is seen 
in several areas.

Figure 3. Bone formation within the tumour. Bony trabeculae with no osteoblas-
tic rimming is seen within the tumour.

Figure 4. The tumor expresses epithelial membrane antigen and progesterone receptor. 
Sections of the tumor were stained with anti-EMA and anti-progesterone receptor primary 
antibodies and counterstained with hematoxylin. (A) Diffuse expression of cytoplasmic 
EMA is seen. (B) Scattered cells show nuclear expression of progesterone receptor.
al surrounded by palisading epithelioid cells. Importantly, published histological features of the calcifying pseudotumor of the neural axis that overlap with those seen in our case are: an interfacing linear pattern of calcification, occa-
sional ossification and EMA expression of the epithelioid cells.\(^{12,13}\) Ossifying fibroma (OF), especially the juvenile psammomatoid (JPOF) variant\(^ {14}\) of the skull is also a differential diag-
nostic possibility. OF is composed of a fibro-
lastic stroma admixed with woven and lamellar bone and basophilic cementum-like material. The proportions of the various components may vary significantly. Furthermore (which adds to the potential differential diagnostic dif-
culties), JPOF characteristically shows osteoblas-
tic rimming that was conspicuously absent in our case. In addition, bone-invading menin-
gioma could mimic a metaplastic meningioma especially in small biopsies and close correla-
tion with biopsy site will be necessary.

The prominent lymphoplasmacytic infiltrat-
ion present in our case gave a vague associa-
tion to the recently described IgG4-related scler-
osing pachymeningitis.\(^ {15}\) In our case, the absence of areas of sclerosis and obstructive phlebitis and the observation that the majority of the cells were lymphocytes (although with focially significant numbers of plasma cells), the predominantly intracerebral location and the results of the immunohistochemical study, strongly militate against this diagnostic possi-
bility. In addition, the patient did not show any clinical features to suggest the possibility of IgG4-related sclerotic disease elsewhere.

In summary, we present a case of a meta-
plastic meningioma with widespread ossifica-
tion and extensive non-psammomatous calcifi-
cation with focal areas forming linear chicken-
wire patterns. The unique pattern of calcification and bone formation has, to the best of our knowledge, not been reported previously and may give rise to a number of interesting, some of which probably just as rarely occurring, differential diagnostic possibilities.

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