Multiple frontal meningioma
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INTRODUCTION
Meningiomas comprise approximately one third of all intracranial tumours. Around 1-10% of all meningiomas are multiple; however, they are not necessarily of the same pathological subtype. The co-existence of typically benign meningioma and an atypical or secretory one is rarely mentioned in the pertinent medical literature.[1-5]

CASE PRESENTATION
A 71-year-old woman was brought by her family to our neurology outpatients’ clinic. The patient had been experiencing progressive cognitive impairment, personality and behavioural changes, and daily dull pancephaline headache for the past few years. Her headache had been poorly responsive to paracetamol tablets. The family said that the patient was irritable most of the time, suspicious, pessimistic, apathetic, and had poor night-time sleep. She preferred social isolation and tried to avoid her neighbours’ visits. Recent memory, registration, and recall were a little bit impaired but her remote memory was intact. Her illiteracy had intersected with several aspects of formal cognitive function assessment. The patient’s vital signs were unremarkable.

Fundoscopy revealed congested optic nerve heads. Bilateral extensor planter reflexes were found. She was neither hypertensive nor diabetic and there was no family history of Alzheimer’s disease or intracranial tumours. The family denied head trauma, domestic abuse, and illicit drug ingestion. There were no features of neurofibromatosis type II and no history of ionizing radiation exposure.

A battery of blood tests was run; complete blood counts, erythrocyte sedimentation rate, urea and electrolytes, liver function tests, random blood sugar, serum vitamin B12, thyroid stimulating hormone, syphilis serology, HIV serology, and general urine examination. All turned out to be within their normal reference range. Chest X-ray, abdominal ultrasonography, and carotid Doppler ultrasonography were unremarkable.

Brain CT scan (figure 1) and brain MRI (figure 2) were ordered. The patient has two supratentorial frontal convexity meningiomas; one on each side. Diffusion...
The patient’s family declined any form of surgical intervention, including biopsy taking. A one-day treatment with intravenous mannitol and a one-week course of dexamethasone did not result in a clinical improvement in terms of cognition and behaviour. Oral zolpidem, on needed basis to help her sleep was prescribed and the patient was given sertraline, 100 mg per day. The patient visited us after 2 months of starting medical therapy. Her behavioural and personality changes improved a little bit according to her family but her cognition was not affected. Zolpidem was used “daily” and it was successful in inducing sleep. The patient died suddenly because of extensive anterior wall myocardial infarction about 3 months later, as her family had stated.

**DISCUSSION**

In 1938, Cushing and Eisenhardt published a landmark book about an intracranial tumour and they named it meningioma.[1] According to Wiemels and colleagues,[2] meningiomas are commonest diagnosed primary intracranial tumours and compromise 33.8% of all primary brain and central nervous system tumours reported in the United States between 2002 and 2006. Several studies found that 2.8% of the general population harbour an asymptomatic intracranial meningioma.[2-4]

Females are afflicted more than males with a male to female ratio of 1:2. The incidence of the tumour increases gradually with age and peaks in the seventh and eighth decades of life.[5] The term multiple meningioma refers to the simultaneous (or sequential) appearance of two (or more) independently located meningiomas, not necessarily of the same pathological subtype and without signs of neurofibromatosis type II.[6-8] The prevalence of multiple meningiomas was thought to be 1-2% of all meningiomas, during the pre-CT era.[1] With the advent of magnetic resonance imaging and its wide use, the incidence has increased to 10%.[6]

A history of exposure to high dose ionizing radiation is the only consistent risk factor for the development of intracranial meningioma. Neurofibromatosis type II substantially increases the risk of meningioma, especially multiple ones.[5] The slight female preponderance and the previous history of breast cancer in some patients may suggest an etiological role of hormonal factors.[9,10] However, Wiemels and colleagues[2] concluded that despite these sentinel clues, meningioma is far from exhibiting a hormone-fed character in the clinic and epidemiologic measures of endogenous and exogenous hormones are not consistently associated with meningioma incidence.

Meningiomas arise from the meningothelial cells of the arachnoid mater and they are commonly found at the supratentorial compartment. These multiple tumours have the tendency towards unilateral hemispheric localization; the cerebral convexity and parasagittal falk are the usual targets. This may be explained by the higher concentration of meningothelial cells at these areas.[1,6]

According to Butti and coworkers, no specific abnormality for multiple meningiomas was found in their study, but their results pointed out the different origin of each tumour and excluded cell migration through the subarachnoid space as a pathogenetic factor in multiple meningiomas.[11] The histopathology of multiple meningiomas does not differ from that of the solitary ones.[12] However, Mocker and co-workers found that the simultaneous occurrence of different grades of malignancy in the nodules is observed in one-third of multiple meningiomas.[13]

The patient’s left meningioma is rounded and displays diffuse homogenous calcification. It has no peritumoural oedema. The appearance is typical of a benign meningioma. The right frontal meningioma is a little bit larger than the left one and shows diffuse and homogenous calcification but the mass is surrounded by prominent peri-tumoural oedema on non-contrast CT brain scanning (figure 1). The oedema extensively involves the right frontal lobe and the anterior part of the right parietal lobe. Filippi and colleagues concluded that the presence of prominent peri-tumoural cerebral oedema usually points out towards a higher grade tumour or a secretory meningioma.[14]

However, Souto and colleagues concluded that two-thirds of their patients with intracranial meningioma demonstrated peri-tumoural oedema and that the site and size of the tumour were probably related to the development of this oedema.[15] They found that peri-tumoural oedema was found frequently in the large meningiomas and that sphenoid wing meningiomas had significantly more peri-tumoural oedema.[6,15]

The patient’s few years’ symptomatology renders the right frontal meningioma unlikely to be highly atypical or malignant meningioma. Secretory meningiomas are rare subtypes which comprise 1.6% of all meningiomas. The male to female ratio is 1:3.[16] However, Regelsberger and co-workers[17] found secretory meningioma in 39 females and 5 males with a female to male ration of 8:1. Their mean age was 58 years. Secretory meningiomas are histologically benign but tend to result in disproportional peri-tumoural oedema, frequently leading to severe medical and neurological complications in postoperative management.[17] Secretory meningiomas contain glandular lumina with secretory globules; these pseudosammmomas are suspected to be the cause of this unusually disproportionate peri-tumoural oedema but the exact mechanism(s) is unknown.[17-19]
Figure 1. Non-contrast CT brain of the patient which shows two right and left-sided frontal convexity meningiomas. Both are rounded and homogenously calcified. The right-sided one is surrounded by prominent peri-tumoural oedema, an appearance which points out towards secretory meningioma; even atypical or frankly malignant meningioma can impart a similar appearance.

Figure 2. Axial T1-weighted MRI with gadolinium. The left frontal tumour took the contrast homogenously and had no surrounding brain oedema. The right frontal tumour enhanced homogenously also but it was surrounded my prominent oedema.

The majority of these secretory meningiomas are found at the frontal convexity.[17,20,21] We depended on the radiological features to diagnose this secretory meningioma. The frontal convexity location and its
hypointensity on T1-weighted MRI, hyperintensity on T2-weighted (and fluid attenuation recover; FLAIR), the bright appearance on gadolinium enhancement, and the prominent surrounding oedema are highly indicative of secretory meningiomas.[22-23]. Because they are slowing growing and benign tumours, the overall prognosis of non-skull base tumours is relatively good and correlates well with the surgical skills and risks and to the extent of tumour removal rather than to the associated brain oedema per se.[22] Partial resected and residual tumour may respond quite well to radiotherapy.

LEARNING POINTS
1. Around 1-10% of all intracranial meningiomas are multiple.
2. Multiple meningiomas are not necessarily of the same pathological subtype.
3. Secretory meningiomas are rare subtypes which comprise 1.6% of all meningiomas.
4. Secretory meningiomas are histologically benign but tend to result in disproportional peri-tumoural oedema.
5. The majority of these secretory meningiomas are found at the frontal convexity.

In summary, our patient had two frontal convexity meningiomas which have resulted in headache, progressive cognitive decline, and behavioural changes. The radiological features are consistent with a typically benign meningioma and a secretory one. Unfortunately, we lack histopathological diagnosis.

REFERENCES
6. Amin OS. Two supratentorial meningiomas: are they different? BMJ Case Reports 2012;10.1136/bcr-2012-007177, Published September 11, 2012.