Mathew *et al.*
Egyptian Journal of Neurosurgery (2024) 39:25
https://doi.org/10.1186/s41984-024-00285-6

Egyptian Journal of Neurosurgery

CASE REPORT Open Access

Frontal ghost tumour: a case report

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Abstract

Background Ghost tumors spontaneously disappear or decrease to less than 70% before definitive diagnosis and treatment (other than steroid treatment). We report our experience with a patient who had not received steroids, and the challenges of managing a ghost tumor from a developing country.

Case presentation A 71 year old female with frontal mass, right proptosis, and frontal headache. Mass was confirmed by cranial CT scan but entirely resolved while the patient was awaiting surgery. Further follow-up at 6 months revealed clinical and MRI evidence of recurrence. Ghost tumors are no myths and can recur!

Conclusion It is imperative to closely follow up with patients who have complete resolution of brain tumors prior to definitive treatment.

Keywords Ghost tumor, Cranium, Vanishing, Case report

Introduction

The clinical diagnosis of a brain tumour comes with many challenges to the patient and even the managing Neurosurgeon as it marks the onset of probably a life-long journey of health care. This becomes more baffling to both parties when the lesion suddenly 'disappears' and cannot be found again. Cases of such experiences have been reported in the literature with various descriptions. Okita et al. in 2012 defined vanishing tumors as tumors exhibiting the radiological feature of spontaneous disappearance or reduction to less than 70% of the initial tumour volume on MRI with Gd-DTPA enhancement before definitive diagnosis and treatment with drugs other than steroids [1]. Numerous causes, notably Central Nervous System (CNS) lymphoma, have been documented [2, 3].

We report a case of a vanishing tumour and the key points we learnt which we believe will be useful in managing these cases, particularly in low-income countries.

Case report

Our patient was a 71 year-old right-handed female who was referred to our neurosurgical outpatient department and presented with frontal swelling and recurrent frontal headache of 3 years duration. The frontal swelling was initially right-sided but increased in size to cross the midline in the last 3 months prior to presentation. No swellings in other parts of the body. She reported no antecedent trauma to the head, no recurrent paranasal infections, and no fever. At about the same time, she noticed recurrent frontal headaches which did not interfere with her daily activities of living initially but worsened within the last 1 month prior to presentation requiring analgesics. No vomiting, seizures, or limb weakness. She was however noticed to have some personality changes characterized by increased aggression, anhedonia, and refusal of feeds. No history suggestive of extra-cranial malignancies. She had no relevant medical co-morbidity of note. Other aspects of history were not contributory. Two weeks prior to presentation, she developed facial pain with occasional blurring of vision which

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necessitated an Ophthalmology review where she also had a cranial CT scan and was referred for neurosurgical care based on CT findings.

At presentation, she was found to be in some painful distress with an otherwise normal general physical finding. Vital signs were within normal limits ($T = 36.3$, Pulse rate = 90 beats/min, Blood Pressure = 120/80 mmHg). She had a GCS of 15 with preserved higher cerebral functions. Cranial nerves functions were preserved. Pupils were 3 mm and briskly reactive to light (direct and consensual) bilaterally. She had normal muscle bulk, tone, power, and deep tendon reflexes across the limbs. Examination of the Head revealed an irregular mass extending from the right frontal area, across the midline extending from the right supra-orbital margin to the bregma with normal overlying skin. No differential warmth and the mass was non-tender. It measured 18 × 14 cm (craniocaudal and lateral dimensions). It was firm, non-mobile, and non-pulsatile. She had no other scalp masses. Examinations of the anterior neck, breast, abdomen, and other systems were clinically normal.

Cranial CT scan showed a hyperdense soft tissue mass with intracranial extension compressing the frontal lobes (Fig. 1a). The lesion showed some enhancement with contrast, with minimal peri-lesional edema. There was sclerosis of the right frontal bone with a moth-eaten appearance and associated spiculations. No paranasal or intracranial collections. Her Fasting Blood Glucose was 4.7 mmol/L. The full Blood Count result was within normal limits. Retroviral screening was negative. Thyroid and abdominopelvic ultrasound scans revealed normal findings.

A diagnosis of Frontal Meningioma to rule out soft tissue tumour (sarcoma) was made. She was placed on analgesics and requested to do cranial MRI and other pre-op investigations. On her pre-surgical assessment visit 4 weeks later, she was unable to do the requested MRI on financial grounds. She however reported resolution of headache and the frontal mass. Clinical evaluation showed minimal right supraorbital swelling. She was able to do a Brain CT scan 3 months later which revealed a normal study (Fig. 1b). The residual right supraorbital

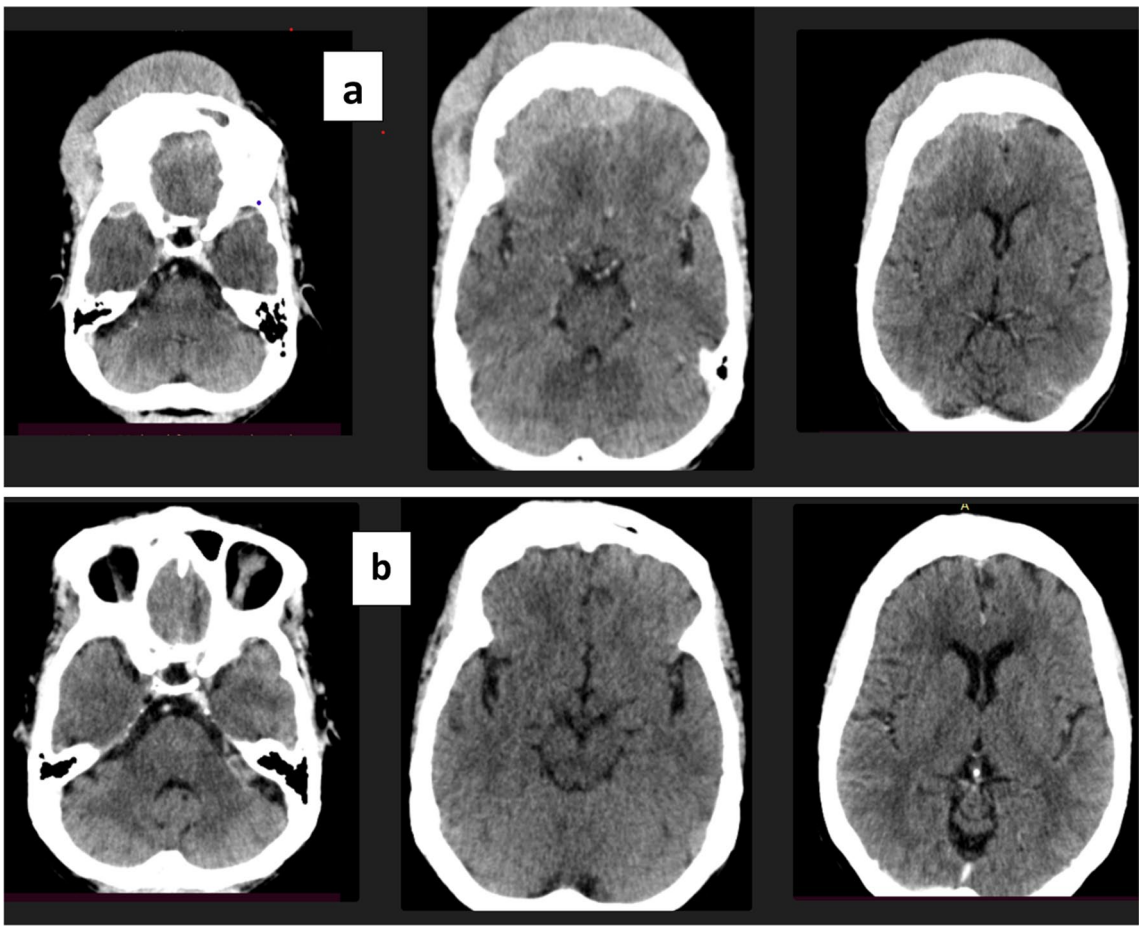

Fig. 1 **a** and **b** Cranial CT scan before and after the disappearance of tumour

mass also resolved completely. However, at 6 months follow up she noticed right supraorbital swelling and mild headaches. A contrasted cranial MRI revealed evidence of recurrence (Fig. 2).

Discussion

Frassanito et al. [3] proposed a classification of Ghost tumors into Vanishing tumors (GhT1), Tumour-like lesions (GhT2), and False tumors (GhT3). According to their classification, a Vanishing tumour describes a space-occupying lesion that shows the radiologic features consistent with the diagnosis of a tumour but unexpectedly disappears or shrinks in size on follow-up radiologic studies, behaving like a ghost! Furthermore, the term spontaneous remission or spontaneous regression (SR) of cancer translates into the recovery of a patient from cancer in the absence of a disease-specific treatment or in the presence of inadequate therapy [4]. While this phenomenon may be the norm in many common ailments, it remains an exception in malignancies hence the concerns when a ghost tumour is encountered in clinical practice.

Various brain conditions have been reported to exhibit this phenomenon. These include, but are not limited to, primary central nervous system lymphoma (PCNSL), gliomas, tumefactive multiple sclerosis, granulomas or tuberculomas, and clival chordoma [5, 6]. PCNSL accounts for up to 50% of cases, but the phenomenon has also been reported with oligoastrocytoma, colonic tumors, renal tumors, etc. [2, 3] When this occurs, the implications could be far-reaching and need to be contained professionally. Patients could understandably question the credibility of the entire clinical process. Therefore, it is important for clinicians managing cancers to be aware of this oncologic phenomenon. In our situation, we did not anticipate it, so we had to discuss it with the patient and her family convincing them that this is a documented phenomenon in clinical practice with a possibility of recurrence. The phenomenon could also come with the tendency for patients to fail to adhere to outlined management protocols. In the typical context of a developing country, the physician might end up being caught in the web of the clinical-supernatural debate. Our patient believed a miracle had happened! Rather than raising objections against the patient's beliefs, we opined that repeat imaging at intervals would go further in ascertaining the miraculous. This was a more noble approach to take than an outright jettison of the patient's faith. In the setting of ghost tumors, this complementary approach will be better suited than a conflicting one.

So why do some tumors vanish? The honest answer is still up for research! Spontaneous tumour regression has been associated with apoptosis, immune system changes, micro-environment, and more or less with DNA oncogenic suppression [7]. Factors that stimulate the immune system have also been postulated and are numerous including bacterial products, enzymes, infections, and hormones [8]. The infection-induced remission of tumors has been proposed. Acute infections are believed to trigger the immune system to eliminate tumors. This has been independently demonstrated with Streptococcus pyogenes [4].

As with our experience, there is a possibility of not having a histological diagnosis in these cases, at least initially. In this patient, the lesion disappeared completely while the patient was awaiting tumour resection as seen in the repeat cranial CT scan (compare Fig. 1a

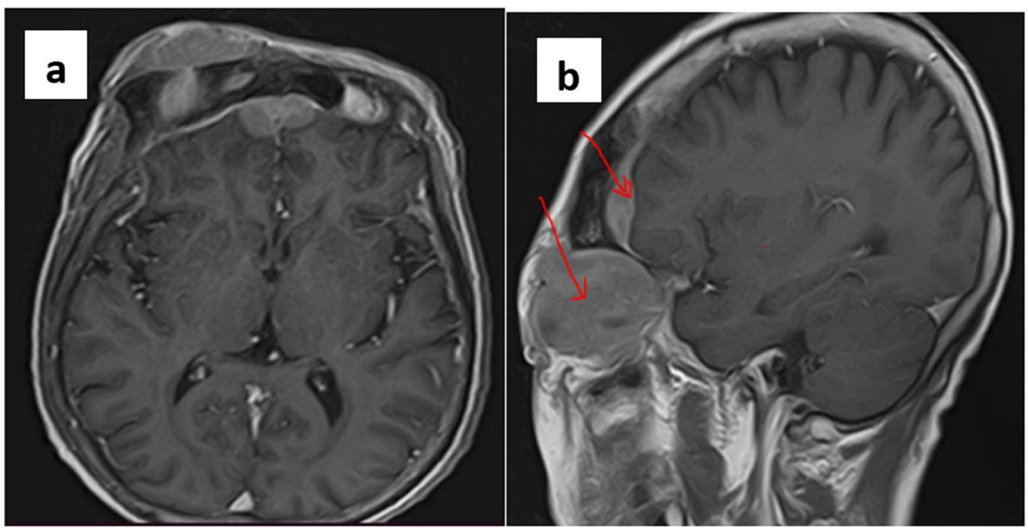

Fig. 2 **a** and **b** MRI Scan at 6 months showing recurrence

and b). Our patient had no prior steroid therapy or any form of intervention specifically directed at the tumour apart from oral analgesics (Paracetamol and Codeine). Such resolution of tumors without intervention has also been reported by Kheiri et al. [9] The use of steroids prior to biopsy has been associated with the resolution of lymphomas in the literature. In a study of 162 PCNSL over a 20-year period in Tokyo, Yim et al. [10] reported a 14% increase in steroid-associated vanishing tumour per 100 mg of dexamethasone equivalent. Sometimes, the resolution of brain lesions can only be picked by imaging. It has been reported that approximately 1% of patients with brain tumors will have disappearing lesions at the time of surgery and because this possibility exists, updated imaging before surgery is recommended [11].

On follow-up, our patient had a recurrence of the right proptosis 6 months after the initial complete disappearance of the tumour, and a follow-up MRI of the brain also showed evidence of intracranial tumour recurrence (Fig. 2). Gupta et al. [12] in their experience, reported a tumour recurrence time of 5 months following the disappearance of a cerebellar cystic medulloblastoma in India. Despite our professional counsel, it has proved difficult for the patient and the family to come to terms and consent to neurosurgical intervention. In a long-term follow-up study of vanishing tumors by Okita et al. [1] in which 10 patients were studied, 5 had a median recurrence duration of 7 months, and they recommended MRI follow-up for all patients with vanishing tumors for up to 5 years. This phenomenon underscores the concepts of individualization of patients and the need for interest in "outliers" whose pathology does not behave as the majority [13].

Conclusions

Ghost tumors are no myths, they do occur! As we go about the business of caring for the sick, it is important that clinicians are aware of this phenomenon. It is also important to follow up on patients whose tumors have vanished using clinical and radiological tools to detect recurrence. As clinicians, our posture must not lean towards antagonizing what has happened, but we should pragmatically walk the patients through the known phases of this poorly understood phenomenon.

Abbreviations

MRI	Magnetic resonance imaging
CT	Computerized tomography
CNS	Central nervous system
GhT	Ghost tumour
SR	Spontaneous regression
PCNSL	Primary CNS lymphoma
DNA	Deoxyribonucleic acid

Acknowledgements

I acknowledge the supportive role played by the son of this patient who was the next of Kin in making this experience to be published.

Author contributions

MM Conceptualization, Design, Drafting, Literature search, and Review of the Manuscript. IG Initial Drafting, Literature search, and Review of the Manuscript. AAA Manuscript Review. SIG Manuscript Review. MRM Review of the final Manuscript. AOJ Manuscript Review.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Availability of data and materials

Not applicable.

Declarations

Ethical approval and consent to participate

Ethical approval was obtained from the Health Research Ethics Committee of the Ahmadu Bello University Teaching Hospital, Zaria with the number ABUTHZ/HREC/F43/2023 for this study. Consent to participate was also obtained.

Consent for publication

Consent for this publication was obtained from the patient for the publication of this case report.

Competing interests

The authors have no competing interests to declare.

Received: 19 August 2023 Accepted: 22 November 2023
Published online: 18 April 2024

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