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Intracranial Teratoma in Young Adult Female: Case Report

Teratoma intracraniano em mulher adulta jovem: Relato de caso

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Abstract

Intracranial teratoma corresponds to a type of germ cell tumor (GCT) of the central nervous system which is characterized by the presence of tissues derived from the germinal layers, which may have hair and adipose tissue inside, for example. The literature on the subject is scarce, and the T2 magnetic resonance imaging (MRI) pattern commonly found is that of a heterogeneously hyperintense mass and, after contrast, teratoma presents heterogeneous enhancement. Therefore, the present case report aims to present atypical results of a GCT in a young adult woman. Data analysis and compilation were performed from medical records, as well as the neuroimaging study. Thus, the present case report demonstrates neuroimaging findings with homogeneous postcontrast enhancement in an adult patient, with an unusual age profile when compared with most of the studies published so far. This demonstrates that it is possible to have atypical neuroimaging findings for mature teratomas and that they can behave in a less aggressive way, not requiring adjuvant postoperative therapies.

Resumo

Teratoma intracraniano corresponde a um tipo de tumor de células germinativas (TCG) do sistema nervoso central que se caracteriza pela presença de tecidos derivados das camadas germinativas que podem conter pelos e tecido adiposo no seu interior, por exemplo. A literatura sobre o assunto é escassa e o padrão de ressonância magnética (RM) T2 comumente encontrado é o de massa heterogeneamente hiperintensa e, após contraste, o teratoma apresenta realce heterogêneo. Pensando nisso, o presente relato de caso tem como objetivo apresentar resultados atípicos de TCG em uma paciente mulher adulta jovem. Para isso, foi realizada a análise e a compilação dos dados em

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- germ cell tumor
- intracranial germ cell tumor
- mature teratoma

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Palavras-chave
- tumor de células germinativas
- tumor intracraniano de células germinativas
- teratoma maduro

prontuário, assim como um estudo de neuroimagem. Assim, o presente relato de caso demonstra achados de neuroimagem com realce homogêneo pós-contraste em indivíduo adulto com perfil etário incomum ao descrito na maioria dos estudos até o momento. Isto demonstra que é possível a confirmação dos achados de neuroimagem atípicos para os teratomas maduros e que eles podem se comportar de forma menos agressiva, dispensando terapias adjuvantes no pós-operatório.

Introduction

Germ cell tumors (GCTs) of the central nervous system are relatively rare, comprising 0.5 to 3% of all intracranial tumors.[1] Germ cell tumors are divided into the following classes: germinomas, nongerminomatous (teratoma, embryonic carcinoma, endodermal sinus tumor, and choriocarcinoma), and mixed GCT. Teratomas are a subtype of GCTs characterized by the presence of somatic tissues derived from two or three germ layers (the ectoderm, the endoderm, and the mesoderm), which commonly affect the child population[2] and, when they affect adults, they are commonly derived from gonadal tissues.[3] They can be divided into mature teratomas, immature teratomas, and teratomas with malignant transformation. Mature teratoma is characterized exclusively by mature adult tissue, while immature teratomas demonstrate components similar to fetal tissue, and teratomas with malignant transformation are very rare and differ because of the malignant transformation of somatic tissue.[4]

Primary intracranial teratomas have a clear male predominance (4:1) and occur predominantly in childhood and adolescence, with only a few cases reported in adulthood.[2,5–7] These tumors tend to appear in the midline structures of the brain, mainly in the pineal and suprasellar regions, possibly due to the great potential of these areas for displacement of progenitor germ cells.[1,8]

Reports of the image pattern of teratomas are scarce in the literature. Most of the knowledge we have are from case reports or from a few small series. Most tumors are solid cystic or predominantly cystic with a mural nodule and show mixed signs derived from different tissues. On T1-weighted images, most lesions show multilocularity or cysts, with or without hyperintensities. On T2-weighted images, most teratomas are shown as heterogeneously hyperintense masses. After the administration of contrast agents, teratomas are usually visualized with a heterogeneous enhancement. But a homogeneous enhancement is possible, as demonstrated in our case.[9,10] With that in mind, the present report aims to present a rare case of CGT in a young adult woman with atypical neuroimaging.

Case Report

The description of the present case report was approved and accepted by the research Ethics Committee (CEP – Plataforma Brasil), under opinion number 4.869.499. The present report refers to a female patient, 36 years old, white, who was admitted to the health system at the emergency service with a complaint of chronic periorbital headache associated with photophobia, phonophobia, nausea, and vomiting – there was a clinical picture of facial paresthesia associated with the headache episodes. According to the patient, she had no previous history of pathologies and did not use continuous medication. There were no complications during the physical and neurological examination.

Through the analysis of neuroimaging by magnetic resonance imaging (MRI) of the skull, there was evidence of an extra-axial mass with expansion to the right middle fossa, with a mass effect in the right temporal lobe. Postcontrast T1 neuroimaging (T1C +) showed a homogeneous hyperintense mass. On the other hand, at T2, the lesion had heterogeneous hyperintensity and diffuse effects of susceptibility, and when at gradient echo (GRE), hypointensity was observed, thus suggesting the existence of calcification or hemorrhage. Therefore, the main initial diagnostic hypothesis was meningioma (►**Figs 1** and **2**).

There was an investigation to exclude the possibility of arteriovenous malformation and cerebral angiography was performed, which indicated absence of vascular and/or flow alterations, discarding the possibility of aneurysm (data not shown). After the complementary examinations, resection of the tumor was performed for further confirmation by anatomopathology. The mass was removed with a size of $\sim 35 \times 30 \times 9$ mm. The resection was performed by right frontotemporal craniotomy and step-by-step debulking using a microsurgical technique. When the tumoral mass was seen during the resection procedure, we instantly identify the presence of a reddish-brown mass with the presence of hairs and heterogeneous-looking histological components (►**Fig. 3**).

After the removal of the tumor, the specimen was sent for histopathological examination, which was confirmed by an anatomopathological report to be a mature teratoma, which contained histological constituents derived from the epidermis, from trabecular bone tissue, and from mature white adipose tissue (►**Fig. 4**).

In the clinical evolution after the surgical period, the patient did not present any neurological deficit and there was a medical record description in which she presented a significant improvement regarding the complaint of headaches. After integrated follow-up with an oncologist, there was no indication of chemotherapy or radiotherapy, and the patient was then submitted to regular radiological follow-up.

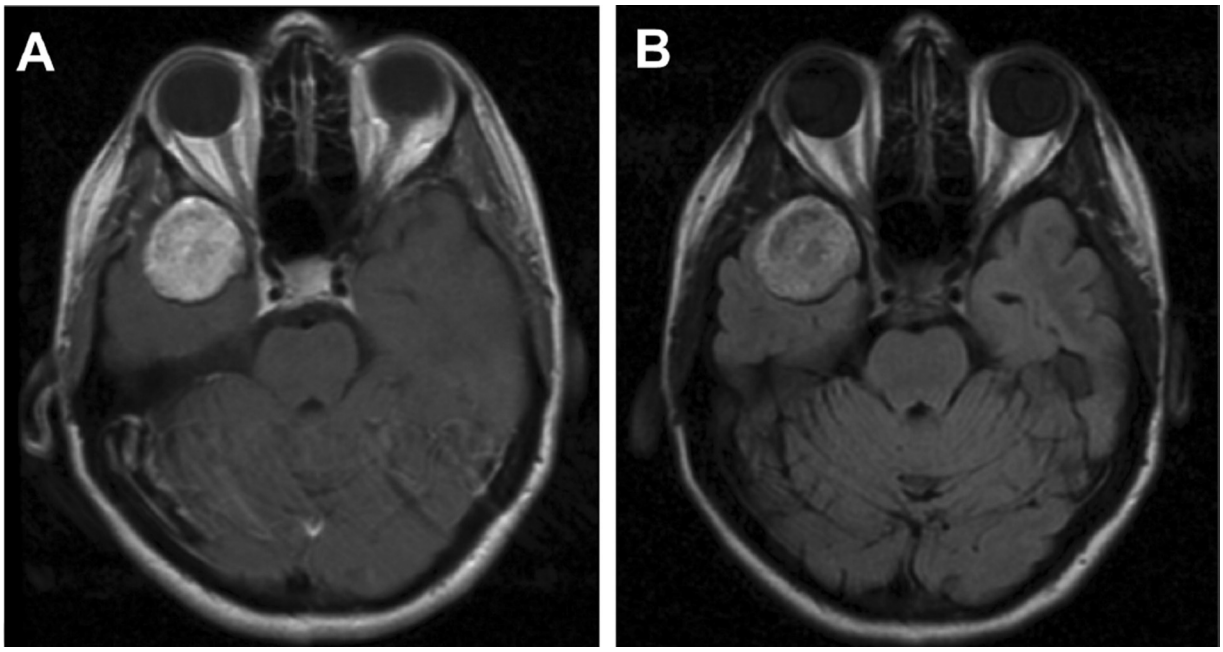

Fig. 1 A. Magnetic resonance imaging in T1C+ – homogeneous and hyperintense mass in the right middle cranial fossa. B. T2 magnetic resonance imaging showing the same mass with a heterogeneous appearance.

Discussion

Mature intracranial teratomas are rare, with 90% of the cases occurring in young individuals < 20 years old.[11] The description of the present case differs from what is most often found in the literature, because in addition to distinguishing from biological aspects for affecting adult women, it also exposes that this extra-axial intracranial teratoma presented atypia in neuroimaging exams.[12–14]

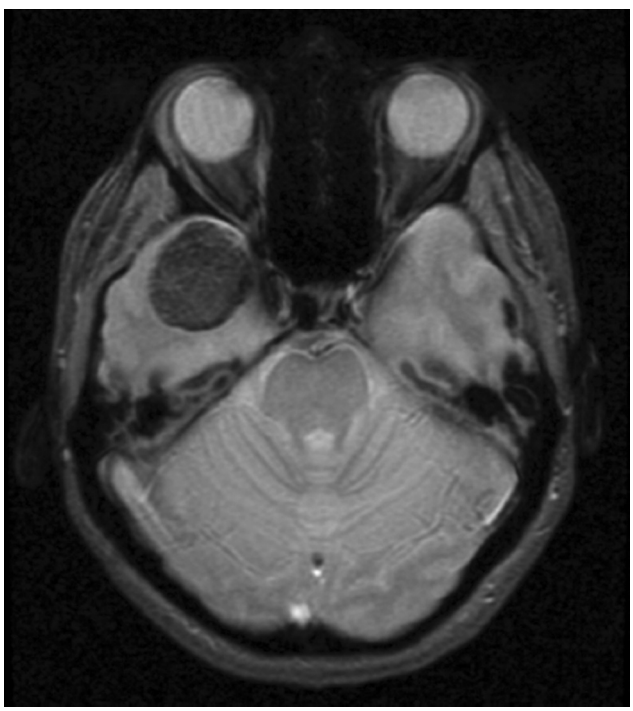

Fig. 2 Gradient echo showing the mass with hypointensity, suggestive of calcification or hemorrhage.

Therefore, knowing that the findings in the literature on the neuroimaging pattern of this type of GCT is scarce, it is recognized that the diagnosis is based on findings in which the results of MRI neuroimaging transmit a mixture of intensity signals in T1 and T2 MRI scans.[15,16] However, in the present case, the results of neuroimaging exams presented as a homogeneous mass in T1C+ and in GRE with hypointensity suggestive of calcification or bleeding. This last finding is identified in approximately half of the cases of mature teratoma, in which there is calcification in its interior[15] as it was also confirmed in the present case through the presence of mature trabecular bone tissue (►**Fig. 4**).

In addition, as expected, according to the histopathological examination, the GCT of the case presented here was also in line with the literature, mostly with tissues derived from the three embryonic leaflets (the endoderm, the mesoderm, and the ectoderm).[14] Specifically, the GCT presented mature tissues such as hair (ectoderm), bone tissue, and white adipose tissue (mesoderm).[17]

As mentioned initially in the discussion of the evolution of the clinical record, the diagnostic hypothesis of an intracranial tumor was mentioned; specifically, it could be of the meningioma type. For this hypothesis, there are clinical findings that lead to such assumption, since, in addition to extra-axial meningiomal formations, MRI showed homogeneous postcontrast characteristics and, also considering that this is the most common primary intracranial tumor with the biological characteristics of the tumor detected in our patient.[18] However, as explained above, the actual diagnosis was different from the initial assumption after the debulking of the tumor *in locu* and the histopathological report.

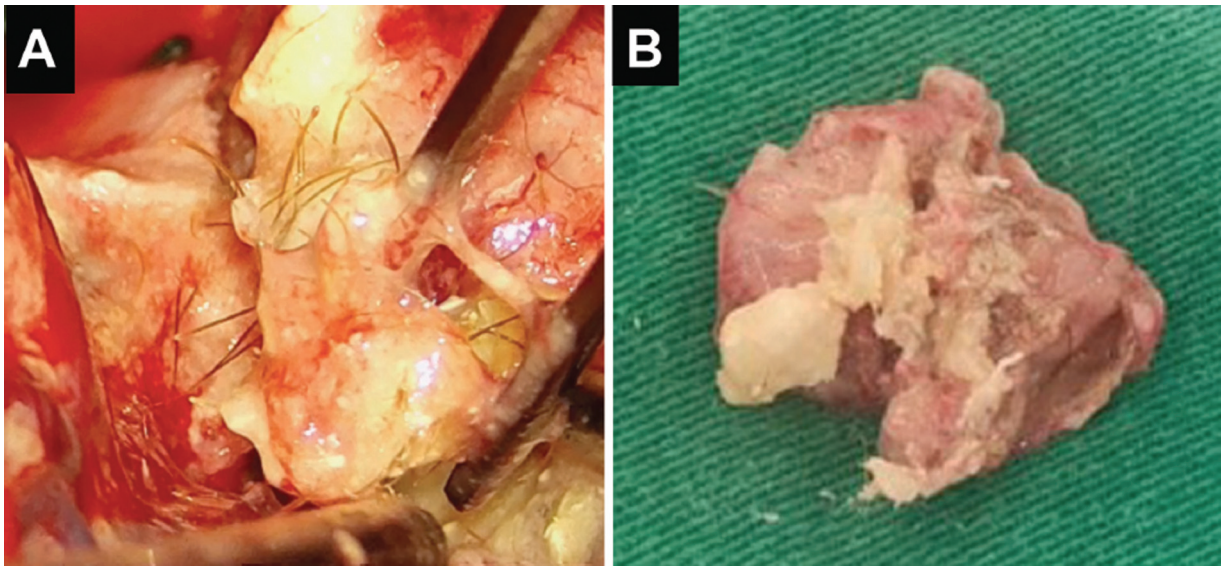

Fig. 3 A. Note the moment of visualization of the tumor by microsurgical technique showing hair strands. B. Tumor mass that was removed, measuring 35 × 30 × 9mm, with a reddish-brown appearance and the possibility of visualizing tissue with histological features similar to those of bone tissue.

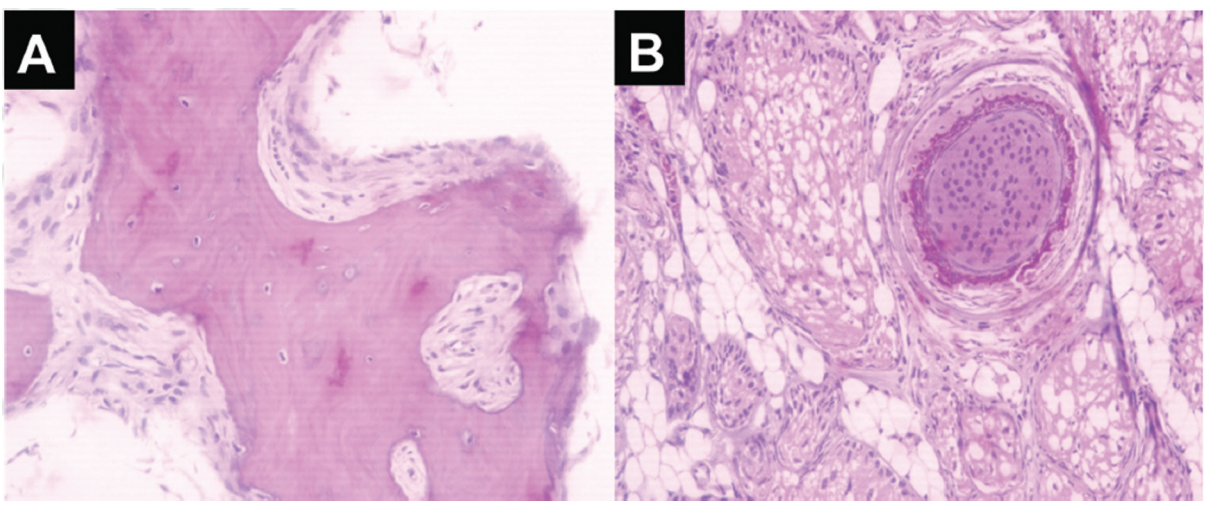

Fig. 4 . View of epidermal and bone tissue in histopathological section. B. Histopathological section showing adipose tissue and trabecular bone.

Regarding the symptomatology of intracranial teratomas, there is no specific and reliable clinical picture, which varies according to the size of the tumor, its location, and course, with findings of intracranial hypertension.[9,19] In the present case report, the manifestation of chronic periorbital headache is justified only by the presence of intracranial hypertension in the patient, as the other symptoms, such as: photophobia; phonophobia; nausea and vomiting together with facial and limb paresthesia were totally unspecific considering the location of the tumor lesion. In addition, nonspecific signs and symptoms are considered since there was no optic nerve compression or intraocular injury, considering that the tumor was located in the anterior portion of the temporal lobe; therefore, far from the posterior occipital portion. Thus, in this scope, one of the possibilities of the symptomatology associated with intracranial pressure would be the existence of a picture of papilledema, as this represents a significant warning sign when there is high intracranial pressure, thus causing not only the potential loss of vision, but also a variety of other visual signs and symptoms such as the ones our patient presented with.[20]

Facial paresthesia may be a finding that is justifiable due to the anxiety of the patient, or it could be explained by compression of the fibers of the cranial nerve root. In both conditions, the literature presents findings in case reports with this sign and symptom; for example, it is known that anxiety can induce hyperventilation, and the resulting hypocapnia and hypocalcemia can cause paresthesia and tetany,[21] just like idiopathic intracranial hypertension.[22]

Generally speaking, CGTs differ in terms of classification and, consequently, in their treatment. In addition to surgical resection of the GCT, options such as chemotherapy and radiotherapy are possible and combinable alternatives.[4] In the present report, the mature teratoma was resected and radiological follow-up was preferentially chosen to optimize

the postoperative period. According to the follow-up of the medical record up to the writing of the present report (~ 12 days after surgery), there was no evidence of recurrence.

Conclusion

This is a case of intracranial teratoma with atypical imaging in a 36-year-old patient. Characteristics such as the age and gender of the patient are uncommon for this type of GCT. Regarding the neuroimaging exams performed in the present case, the homogeneous contrast enhancement – although rare – was a radiological finding and it becomes possible. The treatment of choice presented here was the one most suitable for the case, according to the most accepted protocols, which should preferably be surgical with radiological follow-up, added to chemotherapy and radiotherapy, if necessary, to optimize the postoperative period. With this in mind, we emphasize here that the presentation of a mature teratoma at an advanced age reflects the ideology that these tumors are histologically less aggressive than immature ones.

Ethics

The description of this case report was approved and accepted by the Research Ethics Committee (CEP – Plataforma Brasil), under opinion number 4,727,152.

Conflict of Interests

The authors have no conflict of interests to declare.

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