Intracranial extended Psammomatoid Juvenile Ossifying Fibroma (PJOF)

CASE REPORT & SYSTEMATIC REVIEW

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Abstract

Psammatoid Juvenile Ossifying Fibroma is an uncommon fibro-osseous neoplasm of aggressive but benign nature found in the younger age. Its aggressive path can lead to facial deformation, eye proptosis, and development of intracranial extensions leading to various neurological symptoms. A systematic review based by the MOOSE guideline in Medline, EMBASE and Lilacs, resulted in thirty reported cases of intracranial extended PJOF. Hence, we found it relevant to present a case report of a 15-year-old male with facial deformation and left eye proptosis absent of visual disturbances with PJOF. The lesion was present in the left anterior base of the skull and extended to the intra-orbital space and over the zygomatic arch. The diagnosis was only confirmed as PJOF by histopathological analysis of the completely resected lesion. Our case likes to make awareness of this rare tumor that can be confused with other tumors common to the same topography.

Psammatoid Juvenile Ossifying Fibroma (PJOF) is a very uncommon lesion. It manifests as a benign maxillofacial fibro-osseous neoplasm, which is characterized by the progressive replacement of facial bones with mineralized connective tissues. Subsequently, often resulting in aggressive facial deformation associated to facial pain, swelling, headaches, recurrent sinusitis, nasal obstruction, visual disturbances, or in the worst cases, in blindness [7, 8, 13].

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Keywords

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Our study reports a case of a 15-year-old boy, from the Amazon, with PJOF and considered intracranial extensions, which manifested with left eye proptosis with no visual disturbances and facial deformation. Additionally, our study purposed to list all the medical case reports on intracranial extended PJOF published to date.

Case report

The patient's legal guardian has consented to the submission of the case report to the journal. The Ethics Board of the Hospital Ophir Loyola approved our report.

Chief complaint

A previously healthy 15-year-old male patient natural from Tucumã, a small town in the Amazon, was sent to the neurosurgery department for evaluation of facial deformation on the left side. He is the oldest of three, and the only son of his parents and comes from a low-income family.

Clinical history

The patient manifested a progressive left eye proptosis without any associated pain or visual loss in less than a year. His physical and neurological exams were unremarkable. He was conscious, cooperative, healthy, alert, moderately built and well nourished. His vital signs were all in the normal range. We acknowledged the patient in good health, except for the left eye proptosis.

The eye proptosis was associated with a moderate degree of a left eye displacement and left hemifacial deformity. The skin over the deformed area appeared normal, the temperature in this area was the same as his body's temperature, extrinsic ocular motor movements remained preserved, pupillary reflexes were normal and his visual acuity resulted in 20/20 in both eyes. At left eye fundoscopy we encountered hypervascularity and vascular distensions. His personal and family medical history was unremarkable.

Figure 1: Axial cuts of the magnetic resonance imaging scan in T1 and T2 of the reported case, on which we notice a cystic and multi-lobulated dense lesion in the left frontal area invading the left orbit and para-nasal sinuses.

After noticing an insidious displacement of his left eye, he consulted the local general practitioner, whom requested a Magnetic Resonance Imaging (MRI) scan for further investigation. His MRI scan showed an expansive and large heterogeneous lesion of cystic predominance in the left frontal base of the skull, with multiple lobules and a solid peripheral component, suggestive of osseous density and low aggressive behavior. In **Figure 1**, the MRI scans show a maximum diameter of 75mm x 72mm (line 1 x line 2 in Figure 1) approximately.

Diagnosis

In his first neurosurgical consult, we requested a Computerized Tomography scan (CT-scan) of his

Figure 2: Computerized tomography scans of the reported case taken in sagittal, coronal and axial view, respectively, showing the aggressive extension of the lesion.

brain to see the current state of the lesion. This CT scan (**Figure 2**) presented a large mass of cystic predominance and peripheral solid component. The history, location and progress, suggested a list of possible differential diagnosis.

Therefore, this case was then discussed in the grand neurosurgical rounds, where the board recommended a Digital Subtracted Angiography (DSA) to exclude a possible aneurismal bone cyst. Additionally, the board considered a radical surgical approach despite the DSA results. The DSA came out negative and the patient was submitted for a transcranial surgery, with the purpose to resect the lesion as much as possible, and send a specimen for diagnostic purposes for pathological analysis.

Operation

For the procedure, the patient underwent general anesthesia. After asepsis and placement of sterile surgical cloths, a bi-coronal incision and cutaneous flap dissection was made, followed by a bi-coronal craniotomy to enter the anterior fossa of the skull. By slightly moving the frontal lobe, the lesion could be identified. The lesion was extra-axial, greenish and hard in consistency. The neurosurgeons debulked the lesion from the anterior and middle fossa, the clivus, and the left orbit and among the

ethmoidal bone. This excision resulted afterwards in a large cavity that extended through the above-mentioned structures. The neurosurgeons did not remove the ethmoidal-maxilar extended part of the lesion, which was left to be overviewed and managed by the head and neck surgeon of the hospital. The resected material was directed for further pathology analysis, which confirmed the diagnosis for PJOF (**Figure 3**).

Figure 3: Photomicrograph of the resected material, showing various psammoma bodies, confirming the diagnosis of Psammomatoid Juvenile Ossifying Fibroma (H&E, 100x).

Post-operative course

The patient post-operatively was kept under intensive medical supervision in the surgical intensive care unit for one day. He did not present any surgical or medical complications, showing a quite good post-operative state. On his first post-operative day, he was transferred to the neurosurgical pediatric ward, where he was held for five more days to assure a good clinical reestablishment.

Follow-up

A month after the surgical procedure, the patient did a control CT-scan which showed a complete elimination of the lesion and a repositioned cerebral parenchyma in the vast large cavity where the lesion

used to be. At his follow up consult, the patient did not present any sign of dural lesion or cerebrospinal fluid leaks, neither any other additional complaints. His ocular orbit was less displaced compared to his pre-operative images and the facial deformation showed a slight regression, as expected.

Systematic Review

For the case report we followed the Case Report (CARE) guideline to collect all the necessary data of the patient [5].

For the systematic review of all the reported cases (**Table 1**), we based our record analysis on the

Table 1. Observational systematic review of the reported cases of Psammomatoid Juvenile Ossifying Fibroma with intracranial extensions published in Medline, EMBASE and Lilacs.

Case	Year	Author	Gender	Age	Clinical presentation	Treatment
1	1971	Scott [22]	F	22	Headache and Anosmia	Subtotal Resection. Bifrontal craniotomy
2	1981	Tomita [23]	M	12	Tumor recurrence	Total Resection. Craniofacial approach
3	1983	Villemure [24]	M	22	Swelling, headache and diplopia	Total Resection. Bilateral craniotomy and autogenous Split-Rib cranioplasty.
4	1985	Shields [25]	F	14	Painless, proptosis and upward displacement of the left eye	Total Resection. Bicoronal approach
5	1995	Nakagawa [26]	M	29	Diplopia and exophthalmos	Total Resection. Bicoronal approach
6	1996	Tobey [27]	M	51	Tension pneumoencephalus	Subtotal Resection. Bifrontal craniotomy
7	1997	Lawton [28]	M	34	Progressive loss of vision in left eye.	Total resection. Transfrontal naso-orbital approach.
8			M	36	Recurrent sinus infections and bifrontal headaches. Right eye pain and swelling.	Endoscopic biopsy. Transfrontal-nasal approach. Total resection.
9			M	21	Headache, fever, stiff neck and photophobia.	Endoscopic biopsy. Transfrontal-nasal approach. Total resection.
10			W	22	Left eye swelling and periorbital pain. Headaches	Three sessions of endoscopic resection.
11	1998	Saito [29]	M	15	Meningitis and deterioration of visual acuity.	Total Resection. Bifrontal Craniotomy with Right Temporal Extension.
12	1999	Engelbrecht [30]	M	9	not informed in article	Subtotal resection.
13	2002	El-Mofty [18]	F	53	Nasal obstruction.	Parietal craniotomy.

Case	Year	Author	Gender	Age	Clinical presentation	Treatment	
14	2003	Vandekerckhove [31]	F	16	Displacement of the left globe.	Total Resection. Not described approach.	
15	2005	Baumann [32]	F	41	Facial pain, swelling and exophthalmos.	Total Resection. Transtethmoidal via a Killian incision.	
16	2005	Hasselblatt [33]	M	15	Incidental	Both were totally resected	Lateral rhontomy and a bifrontal craniotomy.
17			M	23	Swelling		Temporal craniotomy.
18	2006	Mehta [34]	M	8	Proptosis of left eye.	Transnasal biopsy	Total Resection. Lateral Rhinotomy and bilateral frontal craniotomy.
			M	11	Proptosis of right eye, epiphora, nasal obstruction and periorbital pain		Total resection through bicoronal craniotomy, extradural removal.
19	2007	Bannykh [35]	F	26	Diplopia, Vertigo and Proptosis.	Biopsy. Trans-sphenoidal biopsy.	
20	2008	Gezici [36]	M	18	Headache and deterioration of left visual acuity.	Subtotal resection. Bifrontal craniotomy.	
21	2009	Nasser [37]	M	12	Left eye proptosis.	Total resection. Left frontal craniotomy.	
22	2009	Noudel [38]	M	12	Seizure, airway obstruction, proptosis.	Total Resection. Bifrontal craniotomy.	
23	2010	Nwizu [2]	F	26	Headache	Subtotal endoscopic resection	
24	2011	Linhares [2]	M	15	Proptosis and downward displacement of left eye.	Follow up first 3 years. Complete surgical removal after progressive worsening.	
25	2011	Tandon [39]	F	?	N/I	N/I	
26	2011	Bohn [12]	M	15	Proptosis and headache	Subtotal Resection. Bifrontal craniotomy and transnasal approach	
27	2012	Geraldo [40]	F	16	Swelling, left eye proptosis, oculomotor dysfunction, reduced visual acuity in left eye.	Total resection. Transcranial approach.	
28	2012	Wehrli [41]	F	N/I	N/I	N/I	
29	2013	Rowland [42]	F	17	Headache and seizures.	Transsphenoidal Biopsy. Initially medical therapy and observed (1.5 years). Surgical approach considered when acute left abducens nerve palsy developed. Endoscopic transnasal approach subtotal resection. Second approach through bifrontal craniotomy for remaining lesion resection.	
30	2014	Liu [43]	N/	I	Headache and intermittent blurry vision in left eye. Nasal congestion and Hyposmia, posteriorly.	Total resection. Endoscopic endonasal approach.	

N/I: means "not informed in article"

MOOSE Guideline [20] for Systematic Reviews and Meta-analysis of Observational Studies in Epidemiology. We conducted the research in EMBASE, Medline and Lilacs databases. The search terms included the association term "AND" with the following terms: "Ossifying", "Juvenile", and "Fibroma". Afterwards additional research was executed, adding the terms "Active", "Aggressive" and "Psammomatoid" or "Psammoma bodies", as this type of tumor has been historically called different names [1]. References in the selected articles were also further analyzed for other papers not captured by the initial literature search.

The found articles were read to filter for case reports regarding Psammomatoid Juvenile Ossifying Fibroma with intracranial extensions. From the selected case reports, we collected data regarding age, gender, clinical presentation, and medical approach for the compilation of the basic epidemiological review of this paper. To avoid any selection bias, two authors separately analyzed the online search and reviewed the articles to be selected.

Discussion

Waldron and Giasanti presented the concepts and criteria of benign fibro-osseous lesions in 1973 after reviewing more than 60 cases [22]. Juvenile Ossifying Fibroma (JOF) has been called many different names since then: juvenile active ossifying fibroma, juvenile aggressive ossifying fibroma and aggressive psammomatoid ossifying fibroma [1]. Because of the confusion, caused by these various denominations, the World Health Organization determined that JOF can only be classified according to their pattern of mineralization in the trabecular (TJOF) or psammomatoid (PJOF) form [19]. These two subtypes vary in their site and age of occurrence, aside their peculiar pathological features [8]. The trabecular subtype usually occurs in the mandible and maxillary bone [17], differently, the psammomatoid subtype usually takes over the or-

bita bones, paranasal sinuses, mandible and has been reported in the anterior intracranial base, part of which the frontal, parietal and temporal bones were involved, but in a very low number of cases [10], causing neurological symptoms and ipsilateral eye displacements [2, 8, 18]. The PJOF variant was at first described in 1938 [4] and is still considered to be very rare (**Table 1**).

PJOF has been diagnosed in any age, but is more incident in males in their second or third decade. This lesion generally presents itself as an asymptomatic progressive rapid expansion of the facial or skull bones [17], causing facial deformation, eye proptosis, diplopia, swelling, hemi-facial sensitivity and pain. In this case report, we presented a patient that had an extended facial deformity and eye proptosis. He presented retro-orbital extensions causing the compression of the ocular globe and local vessels, as noticed in funduscopy.

The diagnosis is currently only determined through pathological analysis of the resected lesion. Microscopic analysis presents a neoplastic histological pattern, which includes the presence of dense cellular fibrous stroma and characteristic spheroidal calcifications called psammoma bodies [2, 18]. Distinctly, these appear with a pink edge and a basophilic center when stained [7], as well as evidence in medical practice, a mass in this topography can be confused with many other lesions. There are case reports that mention the challenging distinction between PJOF, psammomatoid meningiomas in the craniofacial bones, aneurysmal bone cysts and other bone tumors, one mimicking another [1, 2, 12, 21] in the photomicrograph of this case report (**Figure 3**). Recently, a study looking for alternative diagnostic measures blindly analyzed the pre-operative CT scans of patients with TJOF and PJOF. This study assessed the images and compared the finding reported by three experienced radiologists. Interestingly, this study found radiological evidence specific to each subtype [16]. Radiologists report PJOF as an expansile and well-circumscribed radiolucent lesion surround-

ded by a thick wall, as evidenced by this reported case (**Figure 2**).

The aggressive behavior of JOF in general, along with its high rates of recurrence (30-58%) [12] suggests that these lesions should be treated like a local aggressive neoplasm [17], recommending a complete surgical excision with a 5mm safety margin to prevent recurrences [1, 3, 7, 11, 18]. This recurrence can occur in six months or, nineteen years after primary resection, suggesting a long term follow up for these patients and in some situations a re-excision of the lesion [2]. Recurrences can be associated to the difficulty in completing a total resection of the lesion. Some sites and large extensions of the tumor have been reported to be challenging, especially those involving the orbital region, paranasal structures and the base of the skull. In contrary, superficial facial bone tumors are successfully removed in most cases, and present good esthetic results as well. The larger and intracranial extended tumors are usually assessed through craniotomy [6, 14, 15], just like the patient in this case report. In addition, there are also a couple of case reports that mention the endonasal endoscopic [9] approach being also successful in removing TJOF extensions in the orbital cavity [11]. This tumor is contraindicated for radiation therapy as studies reported its radiation-resistant feature [7].

The authors of this article admit some limitations in the reported case, such as the limitation inherent to the retrospective manner of data collection, which might have missed an important detail regarding the case. The short follow-up period after the procedure reported in this paper also is not considerable to define the procedure with a successful outcome, as this is a highly recurrent type of tumor, which in this short time we would not be able to have any signs of such ending. Besides these limitations, all clinical data was carefully searched, collected and analyzed for the compilation of this report.

In this report, we like to emphasize that this is the first PJOF case reported in the northern of Bra-

zil, more specifically from the Amazon region. The difficulty in medical access inherent to the region with a centralized specialized medical care in the capital has influenced in the late diagnosis of this case, which rapidly developed extra and intracranial. Our case report and literature review likes to aware healthcare professionals of this type of tumor, which even though is of benign nature, has the possibility of a very rapid aggressive proliferation, aside the fact that early adequate medical therapy is crucial for a better outcome and avoidance of recurrences.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. This study received no funding to be realized.

Author contributions

Conception and design: Lisboa-Neto. Acquisition of data: Jong-A-Liem, Lisboa-Neto. Analysis and interpretation of data: Jong-A-Liem. Drafting the article: Jong-A-Liem, Nascimento. Critically revising the article: Lisboa-Neto, Nascimento, Paschoal. Reviewed submitted version of manuscript: Jong-A-Liem, Lisboa-Neto, Nascimento, Paschoal, Bor-Seng-Shu, Teixeira. Approved the final version of the manuscript on behalf of all authors: Paschoal. Study supervision: Paschoal.

Note

No portion of this paper has been previously presented or published.

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