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Orbital Compartment Syndrome and Unilateral Amaurosis After Spine Surgery in A Patient with Polyostotic Fibrous Dysplasia

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ABSTRACT

Polyostotic Fibrous dysplasia is a bone disorder showing intramedullary lesions. Orbital compartment syndrome is an uncommon complication of polyostotic Fibrous dysplasia. Patients typically present with acute visual impairment. We describe an 11-year-old male with polyostotic Fibrous dysplasia that developed periorbital bruising and amaurosis affecting the left eye after thoracolumbar arthrodesis. The fundoscopy showed optic disc oedema, a "cherry red" macula and pallor in the posterior pole of the retina. The patient was diagnosed with nonarteritic anterior ischemic optic neuropathy (NAION) and central retinal arterial occlusion (CRAO). The Brain CT delineated the morphological osseous changes of bone, and the MRI demonstrated that the optic nerve was thickened. Polyostotic Fibrous dysplasia can lead to orbital compartment syndrome, especially in a prone position. The long duration of the surgery, the prone position and the pre-existing pathology in this patient were decisive in the disastrous outcome. We suggest intraoperative eye pressure monitoring with decompression before 100 minutes in patients with polyostotic Fibrous dysplasia.

Keywords: Polyostotic fibrous dysplasia; Acute visual impairment; Orbital compartment syndrome; Spine surgery and non-arteritic anterior ischemic optic neuropathy

INTRODUCTION

Polyostotic Fibrous dysplasia (PFD) is a rare genetic bone disorder characterized by the proliferation of immature fibrous tissue in multiple bones; it accounts for 7% of benign bone tumours. Fibrous dysplasia can be monostotic (one bone) or, much less frequently, polyostotic (several bones) and are slightly more frequent in males. The classic radiograph findings are well-defined intramedullary lesions. [1-3] Most of the lesions are benign, while rarely degenerate to malignant forms. Fitzpatrick et al. described in 2004 the typical microscopic findings, which include irregular spindles of woven bone, nonmineralized, scattered throughout a fibrocellular matrix.[4] In general, the occurrence of amaurosis or postoperative vision loss (POVL)after spine surgeries are shallow (< 0,01%).[5,6] Orbital Compartment Syndrome (OCS) is an uncommon complication of PFD that can result in severe visual impairment and require urgent intervention. Although extremely rare, it is associated with significant

morbidity and poses significant challenges to patients and healthcare providers. Several risk factors have been identified, including prolonged surgeries, blood loss, prone positioning, hypotension, anaemia, coagulopathy, use of vasopressors, and direct compression or injury to the optic nerve or vascular supply. Patient-specific factors may also contribute to the risk. The most likely pathophysiological mechanisms underlying POVL after spine surgeries include Nonarteritic Ischemic Optic Neuropathy (NAION), Central Retinal Artery Occlusion (CRAO) and posterior optic neuropathy. Intraoperative factors, such as hemodynamic instability, decreased ocular perfusion pressure, and direct compression of the optic nerve, may contribute to the development of POVL. Patients typically present with visual loss in one or both eyes, ranging from mild visual impairment to complete blindness. Differentiating POVL from other causes of perioperative visual loss is crucial.[4-7]

CASE REPORT

We describe an 11-year-old male with severe PFD (Figure 1). Immediately after thoracolumbar scoliosis arthrodesis, the patient complained of painful acute visual impairment in the left eye, periorbital bruising with proptosis, conjunctival hyperemia, and intense pain on palpation. Standard ophthalmic and ophthalmoscopic examination demonstrated proptosis and periorbital bruising, fixed dilated pupil in the affected eye, complete left eye amaurosis, afferent pupillary defect and restricted extra-ocular muscle movement. The fundoscopy showed optic disc oedema, a "cherry red" macula surrounded by pallor in the posterior pole of the retina with specific periphery areas without signs of retinal detachment. The patient was diagnosed with OCS, presented with NAION and CRAO with these elements. Brain CT and MRI demonstrated that the optic nerve was thickened; there was an increase in soft tissue and proptosis on the left orbit (Figure 2A, B and C).

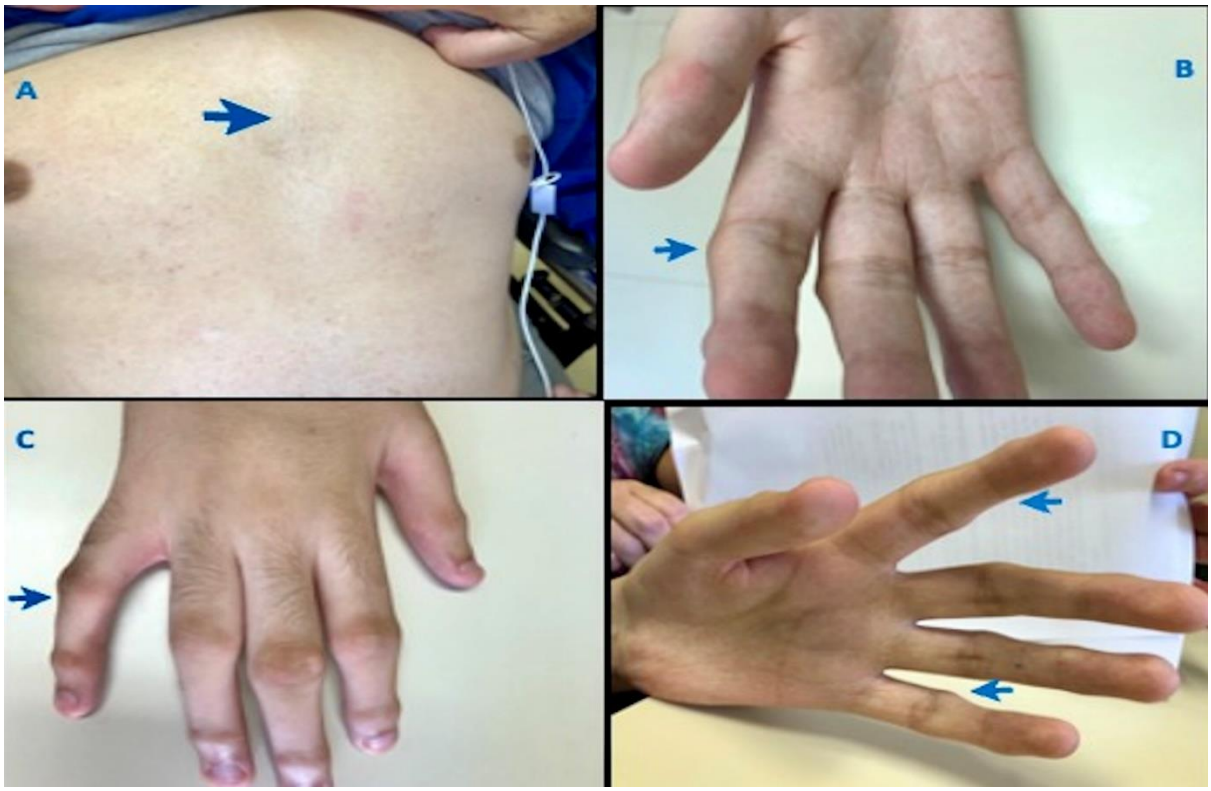

Figure 1: A: The patient presented with normal pigmentation of the skin and Pectus Carinatus (Big Blue Arrow). B, C and D: Noticeble hands deformities caused by polyostotic fibrous dysplasia (Small Blue Arrows).

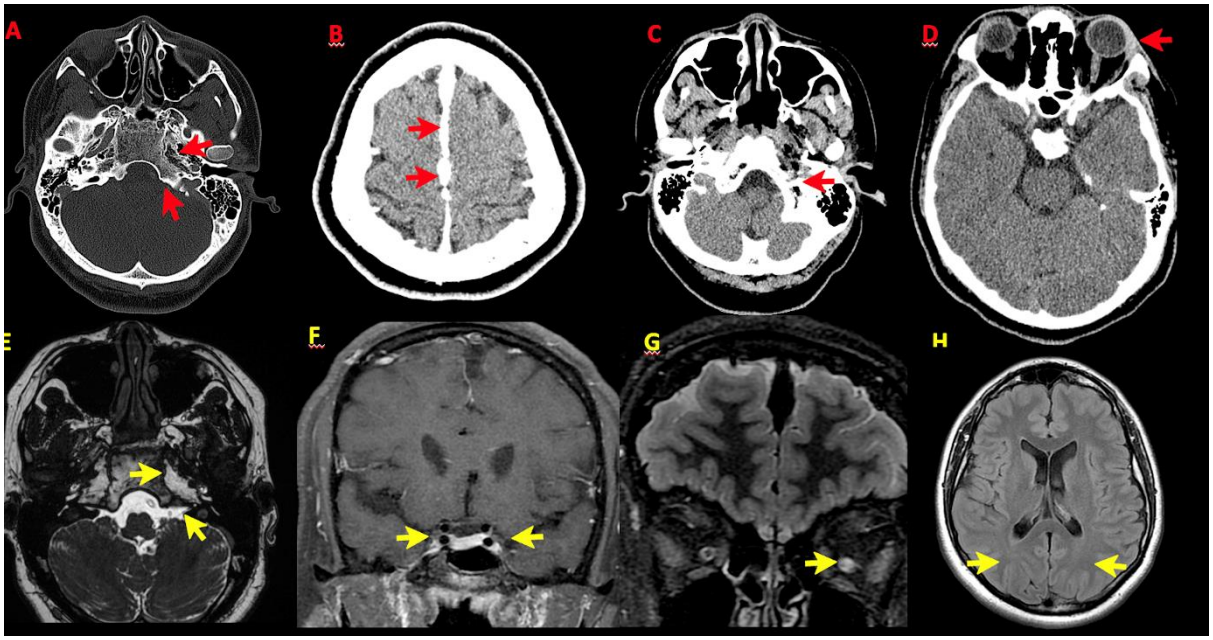

Figure 2: The Brain CT findings. A: The brain CT demonstrates morphological osseous changes in normal skull base anatomy. The osseous part shows irregular curvilinear branching trabeculae of woven bone and the clivus heterogeneously augmented (Red Arrows); B: Exuberant interhemispheric calcifications (Red Arrow); C: Morphological osseous changes in skull base (Red Arrow) and D: Thickening of the left eye's periorbital fat with left eye proptosis. The brain MRI findings: E: The Axial 3D brain MRI shows the presence of a solid expansive process (Yellow left arrows). It involves the clivus with partial obliteration of the sphenoid sinus; F: Coronal T1; the planum sphenoidale; G: Sinals of left optic nerve atrophy and H: Normal brain parenchima. The findings are typical of fibrous dysplasia. The patient fulfilled the Diagnostic criteria according to the WHO classification of soft tissue and bone tumors. [10]

DISCUSSION

Polyostotic fibrous dysplasia is a complex condition affecting multiple bones, including the craniofacial skeleton. Orbital involvement can lead to OCS, a sight-threatening emergency that requires immediate decompression. Diagnosis is often challenging due to the rarity of the condition. Besides ophthalmic examination, brain computed tomography or magnetic resonance imaging is mandatory.[1,2,4]

The patient presented a condition compatible with OCS with central retinal artery occlusion and Non-Arteritic Anterior Ischemic Optic Neuropathy with a poor prognosis. The retina is susceptible to ischemia. In animal models, many authors demonstrated that irreparable damage occurs after 105 minutes of occlusion. The orbital compartment cannot deal with significant increases in intraorbital pressure as the orbit is a closed non-expandable compartment.[8,9] Patients submitted to surgeries in a prone position presenting with AVI must suspect OCS, especially when painful periorbital bruising is present. Managing PFD with OCS requires a multidisciplinary approach involving ophthalmologists, otolaryngologists, and maxillofacial surgeons. The primary goal is to relieve the pressure on the optic nerve and restore visual function. Surgical intervention, such as orbital

decompression and tumour resection, is typically required. Albeit the patient's symptoms started after extubation, the expert opinion was required only 24 hours later; the retinal and optic nerve changes were irreversible. The long duration of the surgery, the prone position and the pre-existing pathology in this patient (clivus tumour with sphenoid sinus obliteration) were decisive in the disastrous outcome (Figure 2).

Assistant doctors should know this potential complication in patients with fibrous dysplasia presenting with proptosis, visual disturbances, or facial asymmetry. We suggest intraoperative eye pressure monitoring (hand-held tonometer) with immediate decompression in less than 100 minutes for high-risk patients.[4]

REFERENCES

1. W H Jee, K H Choi, B Y Choe, J M Park, K S Shinn. Fibrous dysplasia: MR imaging characteristics with radiopathologic correlation. AJR Am J Roentgenol. 1996;167(6):1523-7.
2. Resnick D. Diagnosis of bone and joint disorders, 4th ed. Philadelphia, PA: Saunders, 2002:4285 –4840.
3. Joyce KM, O'hEireamhoin S, Sheng MT, Devitt A. Fibrous Dysplasia of the Spine - A Case Involving Three Levels of Thoracic Spine. J Orthop Case Rep. 2014;4(2):73-77.
4. Fitzpatrick K, Taljanovic M, Speer D, Anna R Graham, Jon A Jacobson, George R Barnes, et al. Imaging Findings of Fibrous Dysplasia with Histopathologic and Intraoperative Correlation. Am J Roentgenol. 2004;182(6):1389-1398.
5. Zimmerer S, Koehler M, Turtschi S, Palmowski-Wolfe A, Girard T. Amaurosis after spine surgery: survey of the literature and discussion of one case. Eur Spine J. 2011;20(2):171-176.
6. Patil CG, Lad EM, Lad SP, Ho C, Boakye M. Visual loss after spine surgery: a population-based study. Spine Phila Pa. Spine (Phila Pa 1976). 2008;33(13):1491-6.
7. Berry S, Lin WV, Sadaka A, Lee AG. Nonarteritic anterior ischemic optic neuropathy: cause, effect, and management. Eye Brain. 2017;9:23-28.
8. Hayreh SS, Kolder HE, Weingeist TA. Central retinal artery occlusion and retinal tolerance time. Ophthalmology. 1980;87(1):75-78.
9. Ray-Chaudhuri N, Kiné DA, Tijani SO, Parums DV, Cartlidge N, Strong NP, et al. Effect of prior steroid treatment on temporal artery biopsy findings in giant cell arteritis. Br J Ophthalmol. 2002;1;86(5):530-2.
10. WHO Classification of Tumours Editorial Board. WHO Classification of Tumours, 5th Edition. Soft Tissue and Bone Tumours. (2020) ISBN: 9789283245025 - IARC Publications.