CASE REPORTS

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# Central retinal artery occlusion following embolization in juvenile nasopharyngeal angiofibroma – A case report

## Okluzija centralne arterije retine posle embolizacije juvenilnog nazofaringealnog angiofibroma

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**Abstract**

**Introduction.** Juvenile nasopharyngeal angiofibromas are highly vascular, locally aggressive lesions, that affect male adolescents. The surgery is the treatment of choice, although it shows a strong propensity to bleed during surgical removal. Preoperative embolization enables the surgical approach in a less bloody way and also a complete resection of the tumor. However, this procedure is not without complications. The most severe complication of this technique is a migration of an embolus into the intracranial circulation. **Case report.** We present a 9-year-old boy who lost vison on his left eye following preoperative embolisation of juvenile nasopharyngeal angiofibromas as a result of central retinal artery occlusion. A recent review of the literature reported only three previously documented cases of central retinal artery occlusion occurring after embolization for a nasopharyngeal angiofibroma. We want to point out the possibility of this rare but devastating complication and the importance of rapid and accurate diagnosis and treatment so that a visual outcome could be better when applying an early medical treatment. **Conclusion.** Described case of central retinal artery occlusion is a rare and unusual, iatrogenic vascular event, that could arise as a complication from embolisation of nasopharingeal tumors. However, physicians (ophthalmologists and ear-nose-throat surgeons) should be aware od this devastating complication, and the close evaluation of angiograms for detection of any vascular abnormality before and during the embolization is crucial.

**Key words:**
**nasopharyngeal neoplasms; angiofibroma; child; diagnosis; embolization, therapeutic; otorhinolaryngologic surgical procedures; neoplasm recurrence, local; retinal artery occlusion; treatment outcome.**

**Apstrakt**

**Uvod.** Juvenilni nazofaringealni angiofibrom je benigna, vaskularizovana, lokalno agresivna lezija, koja se obično javlja kod adolescentnih momaka. Terapija izbora je hirurško uklanjanje. Kako ovaj tumor pokazuje sklonost ka krvarenju tokom hirurškog uklanjanja, uvođenjem preoperativne embolizacije omogućuje se operacija sa manje krvi, ali i kompletna resekcija tumora. Međutim, ova procedura nije bez komplikacija. Najozbiljnija komplikacija embolizacije jeste migracija embolusa u intrakranijalnu cirkulaciju. **Prikaz bolesnika.** Prikazujemo slučaj devetogodišnjeg dečaka koji je izgubio vid na levom oku posle preoperativne embolizacije juvenilnog nazofaringealnog angiofibroma zbog okluzije centralne arterije retine. Pregledom literature našli smo svega tri objavljena slučaja okluzije centralne arterije retine nakon embolizacije nazofaringealnog angiofibroma. Ovim radom želimo da ukažemo na mogućnost ove retke i teške komplikacije, kao i na značaj brze i tačne dijagnoze, jer se bolji vidni ishod dobija kod ranog započinjanja lečenja. **Zaključak.** Opisani slučaj okluzije centralne arterije retine je redak, jatrogeni vaskularni akcident koji može da se javi kao komplikacija embolizacije nazofaringealnih tumora. Lekari (oftalmolozi i otorinolaringolozi) bi trebalo da budu svesni ove teške komplikacije, kao i toga da je ključno detaljno praćenje angiograma radi detekcije bilo kakve vaskularne anomalije, pre i tokom embolizacije.

**Ključne reči:**
**nazofarinks, neoplazme; angiofibrom; deca; dijagnoza; embolizacija, terapijska; hirurgija, otorinolaringološka, procedure; neoplazme, lokalni recidiv; okluzija retinalne arterije; lečenje, ishod.**

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## Introduction

Juvenile nasopharyngeal angiofibromas (JNA) are highly vascular, benign but locally aggressive tumor-like lesions, that commonly affect adolescents [1]. The origin of JNA are still uncertain concerning its fibrous or vascular derivation. While some authors considered JNA as vasoproliferative malformation because of its extensive vascularisation, the others proposed JNA as a specific type of hemangioma [2]. Recent evidence based on immunohistochemical and electron microscopic examinations indicates that JNA represent vascular malformations derived from incomplete regression of the artery of the first branchial arch, rather than a true neoplasm. Nevertheless, the exact etiology of these lesions remains a matter of debate [3].

Traditionaly, the surgery is considered as a treatment of choice. However, because of its rich vascularisation, JNA shows a strong propensity to bleed during surgical removal [4]. The introduction of endoscopes and preoperative embolization of the feeding vessels, changed the surgical approach of these tumors by providing an operation in a less bloody way and complete resection of the tumor [5]. However, embolisation is not a procedure without complications. The most fearing complication would be migration of an embolus into the intracranial circulation, while the other complications include systemic reaction to the contrast, infection at the site of puncture, femoral hematoma and thrombosis, facial paralysis, skin necrosis, oronasal fistula [6].

We present a 9-year-old boy who lost his vison on his left eye following preoperative embolisation of JNA, as a result of central retinal artery occlusion (CRAO). Recent review of the literature reported only three previously documented cases of CRAO occurring after embolization for a nasopharyngeal angiofibroma [7–9]. We want to point out the possibility of this rare but devastating complication and the importance of rapid and accurate diagnosis and treatment so that a visual outcome could be better when applying an early medical treatment.

## Case report

A 9-year-old boy was presented to the Ear-Nose-Throat (ENT) Clinic with a complaint of left nasal obstruction, repeated left-sided nose bleeds and snoring of few months duration. On the clinical examination, purplish mass filling the left nasal cavity was found. The triad of epistaxis, one-sided nasal obstruction and a mass in a nasopharinx are indicative for JNA so the diagnosis of this tumor was proposed. Computed tomography (CT) showed a heterodense, soft tissue lesion in the nasal cavity, with measurement 35 × 40 × 58 mm, displacing the nasal septum and extending partially to the sphenoid sinus. It was recommended to embolize the feeding vessels of the tumor using polyvinyl alcohol (PVA) particles as a part of preoperative preparation in order to reduce the size of the tumor and to reduce the possibility of bleeding during the surgery. Five days after the embolization process, the patient underwent an endoscopic angiofibroma excision. The histopathologic finding confirmed the JNA. Eight months after the operation, patient is presented with reccurence of the symptomatology. On control CT scan, the hyperdense mass filling the nasal cavity, measuring 17 × 27 × 16 mm was found. Comparing the clinical and imaging findings, a diagnosis of residual JNA was made. Endovascular angiography of the both carotid and vertebrobasillar system was carried out. At angiography, it was noticed that most of the blood supply to the lesion arises from the maxillar artery, so this artery was then embolised (Figure 1).

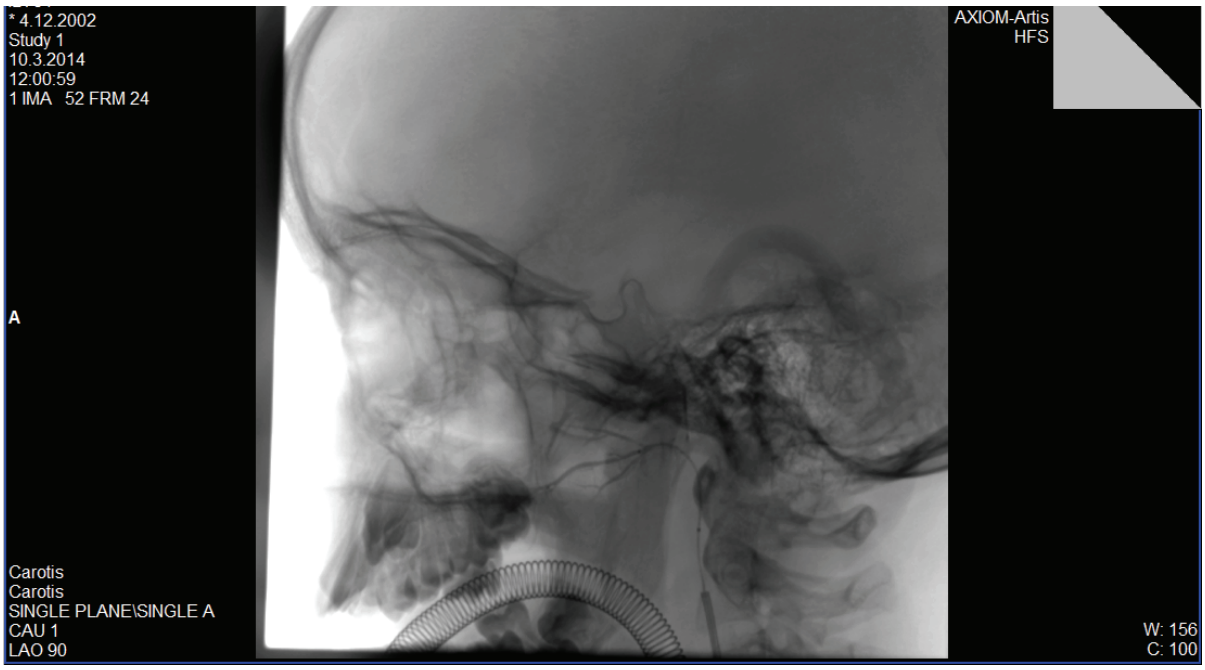


**Fig. 1 – Angiogram of the external carotid vascular system showing the vascular angiofibroma. Note the appearance of a tumor blush.**

Following the procedure, the patient awoke but with impaired conscious, somnolent, with highly positive meningeal signs such as stiff neck, positive Brudzinski and Kernig sign. His left pupil was dilated and nonresponsive to light. As the patient was somnolent and desoriented, he was examined by a neurologist, and it was performed an urgent CT scan and brain magnetic resonance imaging (MRI) and angiography (MRA). The CT scan showed the left hemisphere edema, while MRI showed microischemic lession and left subarachnoid front-parietal hemorrhage. Prompt treatment with antibiotics and antiedematous therapy with mannitol and dexamethasone was started. He was also examined by an ophthalmologist the following day after the vascular event. Left eye visual acuity (VA) was not perceptive of light and the relative afferent pupillary defect was positive in this eye. The patient's fundus examination showed whitish retinal edema and a cherry-red spot appearance of the macula with narrowed vessels (Figure 2).

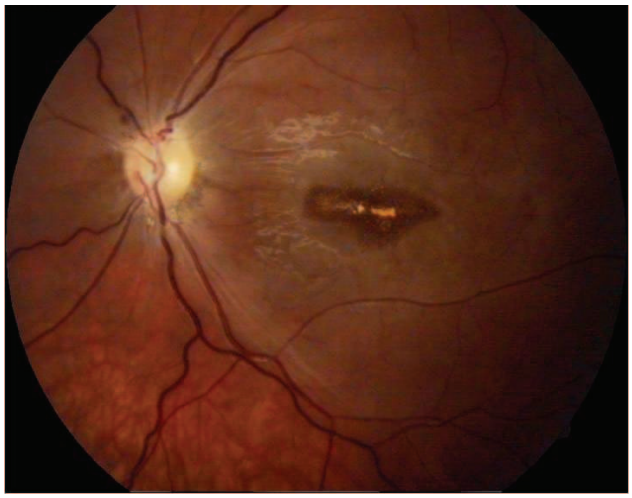

**Fig. 2 – Fundus changes on the left eye one months after the vascular event. Note maintained whitish appearance of the macula with cherry-red spot and narrowed vessels.**

A diagnosis of central retinal artery occlusion was made. Ocular massage was initiated and proceeded for 15 minutes. Topical timolol was given twice a day. The patient's general condition improved slowly with progressive resolution of neurological signs and partial resorption of cerebral edema on the brain CT scan. On the other hand, the visual loss on the left eye was still persistant. Two month after the vascular incident, the patient was conscious with no neurological deficit. The only problem he had was low vision on the left eye. After detailed discussion with the parents, it was decided to perform an operation again. Five days after the repeated embolization process, the patient underwent endoscopic angiofibroma excision and the tumor was removed completely. Ten months after the embolization, VA was still no light perception. Fundus changes on the left eye such as cherry-red spot disappeared while it was noted attenuated retinal arterioles and optic disc atrophy (Figure 3).

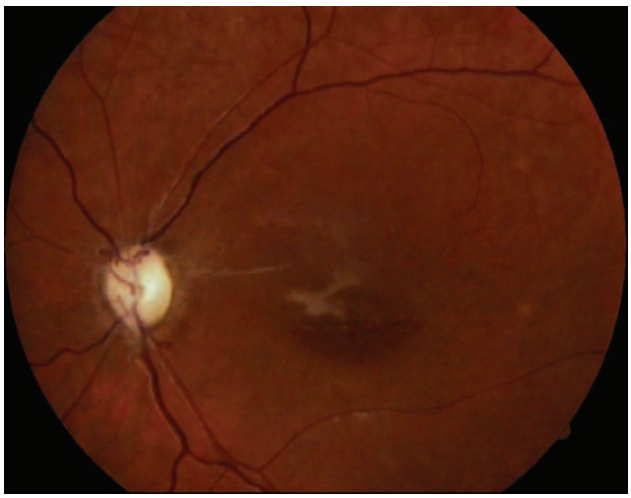

**Fig. 3 – Fundus changes on the left eye three months after the vascular event. Note attenuated retinal arterioles, optic disc atrophy and dissapearance of cherry red spot.**

## Discussion

JNA represents the most often head and neck vascular malformation in males in pre-puberty period [2, 9]. Symptoms that occur are typical for JNA: progressive unilateral nasal obstruction accompanied with rhinorrhea and recurrent epistaxis. Depending of tumor extension to surrounding structures, rhinosinusitis, alteration in olfaction, proptosis, vison alteration, headache and neurologic deficit are also possible manifestation [10].

Although different treatment modalities are used for angiofibromas such as surgery, hormonal therapy, radiation and systemic chemotherapy, a surgical excision of the mass remains a treatment of choice [11]. As the JNA is highly vascularised tumor, blood loss during surgical resection is one of the major preoccupations during the operation. The appropriate surgical approach should be determined by performing the preoperative transcatheter embolisation of the tumor [2] in an attempt to decrease intraoperative bleeding and to make tumor resection more easy [12]. Because of the often bilateral vascular supply, both carotid systems should be angiographically evaluated [2]. However, preoperative embolisation is not without complications. The most severe complications, like cerebral infarcts and vision loss, have an incidence of less than 2% [5]. Central retinal artery occlusion and subsequent vision loss, as seen in our case, were reported only few times in literature after JNA embolization [7–9].

There are three mechanisms describing in which way PVA embolisation cause CRAO: the congenital variation of vasculature, over-forced injection which cause a reflux into the internal carotid system and presence of collateral vessels which arise from tumor [9]. The presence of collateral vessels could be masked by a tumor itself, but if it is recognized, the microcatheter should be advanced beyond the second portion of ophthalmic artery to prevent the embolic event [1].

Ramezani et al. [7] reported a case of a child with right sided JNA who developed CRAO following preoperative embolisation, probably due to the presence of suspicious collateral artery between the external carotid artery and oph-

thalmic vessels on the left side which had not been noticed before the embolisation. On the other hand, Casasco et al. [4] assumed that, in their case, a small amount of permanent liquid polymerizing agent entered the ophthalmic artery, which resulted in an acute loss of vision due to CRAO.

In our case, we did not find any vascular abnormal communication nor collateral which could explain the route of the embolus which affected the ocular circulation. We could assumed that the changes were caused by the PVA material pass through the cerebral circulation and internal carotid artery and its branches supplying the retina.

There are some authors who found the preoperative embolisation as a risk factor, with a higher rate of JNA recurrence [13, 14]. Petruson et al. [13] found that the recurrence rate in non-embolized patients was 8% and 41% among embolized patients. In their opinion, the only factor affecting recurrence was the age at the moment of making a diagnosis, i.e., the younger the patient was, the greater the risk for recurrence. Yet, they hope that the development of imaging and embolization techniques will contribute to reducing the recurrence rate. On the other hand, Ogawa et al. [6] found the embolisation as an effective technique for decreasing the tumor size and easier way for resecting it, thus lowering the recurrence rate of the tumor. They concluded it after reviewing the medical records of 170 patients who underwent preoperative embolisation for resection of JNA, confirming that recent development of embolization techniques, made embolisation even safer and more effective.

Nevertheless, this emergency vascular accident continues to be an undesirable and tragic event for anyone affected by it, especially because of the difficulties in preventing and managing CRAO [11].

## Conclusion

Described case of CRAO is a rare and unusual, iatrogenic vascular event, reported only few times in literature (according to PubMed search), that could arise as a complication from PVA embolisation of nasopharingeal tumors. However, physicians (ophthalmologists and ENT surgeons) should be aware od this devastating complication and the close evaluation of angiograms for detection of any abnormality before and during the embolization is crucial. Since, visual prognosis would be much better with applying an early treatment, it is extremely important to set rapid and accurate diagnosis of CRAO and to treat all such patients within a few hours after the occlusion. On the other hand, all patients undergoing these procedures (or their parents) should be fully explained and informed about the risk of visual loss as it could strongly influence their future quality of life.

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