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Case Report Open Access

Juvenile Angiofibroma Occurring in an Unusual Site: Report of a Case

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Abstract

Extra Nosopharyngeal Angiofibromas (ENA) is a tumor of fibrotic and angiovascular consistency which are localized outside the nasopharyngx (i.e. nasal septum). The clinical characteristic of ENA and JNA (juvenile angiofiborma) is not conclusive and the diagnosis of the ENA is problematic. The nine year old girl was administered to the Children Otolaryngology Clinic. The histopathological examination proved that the removed mass was angiofibroma. According to literature and our clinical experience it is important to be aware of the atypical localization of the juvenile angiofibromas. According to literature only about 15 cases of the similar case were reported.

Keywords: Angiofibroma; Tumour; Cancer; Epistaxis; Female

Introduction

Angiofibromas are rare, benign tumors of the vascular and fibrous connective tissue composure. Although the lesion is benign it can present local malignancy. In advanced cases it can extend intracranially. The lesion was first recorded by Chelins in 1847 and the name "angiofibroma" was given by Friedberg in 1940 [1]. JNA (juvenile angiofiborma), appear almost exclusively in adolescent males between 14 and 25 years [2]. Only about fifteen cases of the angiofibroma in adolescent females were described. The incidence of the JNA is less than 0.5% of tumors arising in the head and neck [3]. In USA this lesion is the most frequent head and neck tumor of the adolescence with 1 new case per 5000 to 50000 patients referred to otolaryngologist [4]. Glad and colleagues reported an incidence of JNA in Denmark of 0.4 cases per million inhabitants per year. In the Middle East and India the incidence seems to be much higher than in Europe. Symptoms of the JNA depend on tumor size, localization andextend. The epistaxis and the nasal obstruction are the most common [5] but due to tumor extrapharyngeal extension different symptoms can be reported and these are rhinorrhea, smell alterations, hypoacusis, facial swelling, otitis media [2].

Background and Case Presentation

We describe the case of a nine-year-old-girl which was administered to the Emergency Ward of the Children Clinical Hospital in Lublin. The most concerning symptom was epistaxis. The child was consulted with otolaryngologist. Nasal cavity was cleaned from the blood clots and in the anterior rhinoscopy the bleeding mass of about 1 × 0.5 cm was discovered. It was situated on the left side of the nasal septum. The nasal septum is a very rare site of extra nasopharyngeal angiofibroma with only two cases reported in the medical literature [6]. The laboratory tests did not present any abnormalities both the blood morphology and the coagulation parameters were normal. The patient was consulted with the oncologist and the decision about the operation and the histopathological examination was made. The procedure was performed in general anesthesia, the throat was previously protected with the gauze package, under the control of the endoscope 0° using knife, the circumferential cut was made and lesion was totally removed and the margin was preserved. The bleeding was equipped with coagulation and sutures. The material was send to the histopathological examination. Histologically this tumor is composed of two basic components: fibrocallagenousstroma and many vascular channels. The vascular component is made up of thin-walled, small to medium-sized vessels and scattered thick-walled vascular channels. The stroma is composed of fine or coarse collagen fibers. The stromal cells are spindle shaped. Long-standing lesions may show stromal hyalinization. Mitotic figures are rare and usually no cytologicatypia is seen (Figures 1-7).

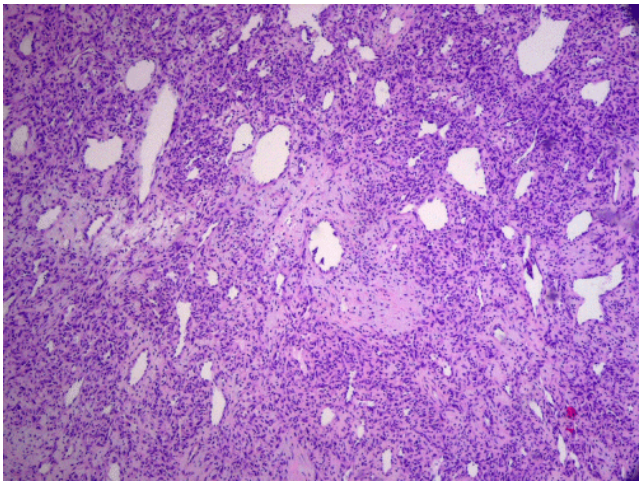

Figure 1: Angiofibroma. Note the numerous variably sized vessels focally associated with collagen fibers. H+E.

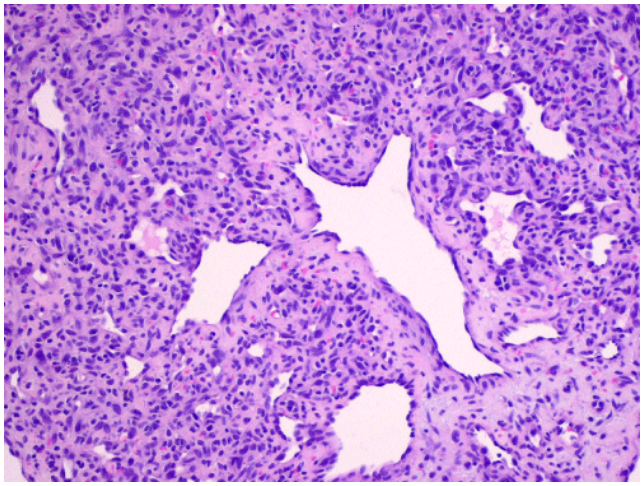

Figure 2: Irregular vascular channels in angiofibroma. Note the prominent capillary –sized vessels. H+E.

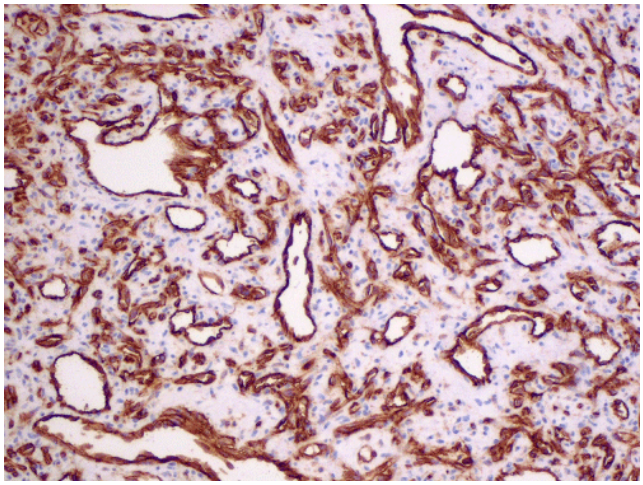

Figure 3: Immunostain for CD34 in angiofibroma.

Radiographic Features

The computed tomography (Figure 7) and the magnetic resonance are the mainstay in the case of ENA (extranosopharyngeal) because the biopsy is contraindicated. In our case the computed tomography examination was performed. On the left side of the nasal septum on its upper edge the pathological area was discovered (Figure 8 and 9). Its density was about 93 jH with enhancing to 143 jH (Figures 1-3). The lesion was well limited of the size of 12 × 8 mm, it partially blocked the left nasal duct (Figure 10). Sinuses were free from changes. In the chest X-ray no changes were found.

Discussion

A lot of theories exist about the origin of the juvenile angiofibroma. In one of the hypothesis JNA is considered to be a vascular malformation. It was considered to be a specific type of hemangioma or ectopic proliferation of the vascular tissue. More recent studies suggest that JNA is more a vascular malformation (or hamartoma) than tumor [4]. Schick et al. postulate that the lesion can arise due to incomplete regression of the first branchial artery. Many authors underline the role of the androgen and/or estrogen receptors in the tumors development however hormonal pathogenesis of JNA is still unclear. Also the hormonal treatment is ineffective. Many studies proved the numerous chromosomal alterations among patients with JNA. It also have been proved that patients with familial adenomatous polyposis (FAP) have increased prevalence of JNA [4].

Our case presents an example of extranasopharyngeal angiofibroma. Such lesions most commonly originate from maxillary sinus. Other localizations are the ethmoid and sphenoid sinuses, nasal septum, middle turbinate, inferior turbinates, conjunctiva, molar and retromolar region, tonsils and larynx. ENA are more frequent among females at later age [7].

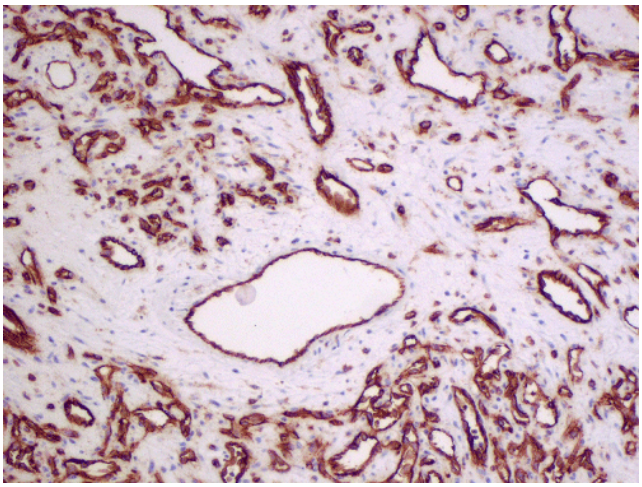

Figure 4: Strong, diffuse positive immunoreactivity in endothelial tumor cells.

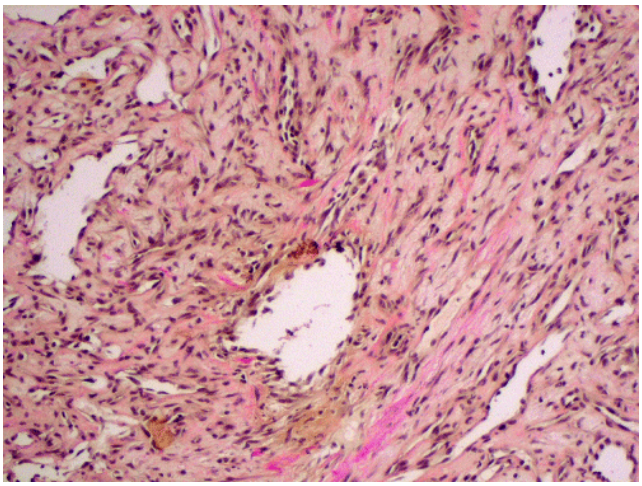

Figure 5: The stroma in angiofibroma with van gieson stain.

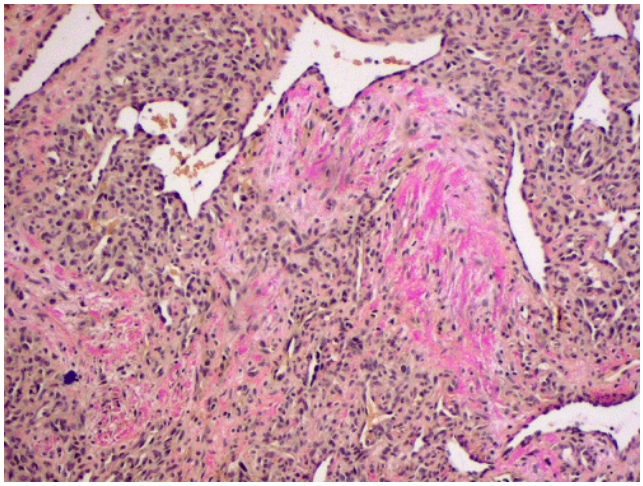

Figure 6: Wispy collagen fibers arranged in short fascicles and surrounding vascular channels.

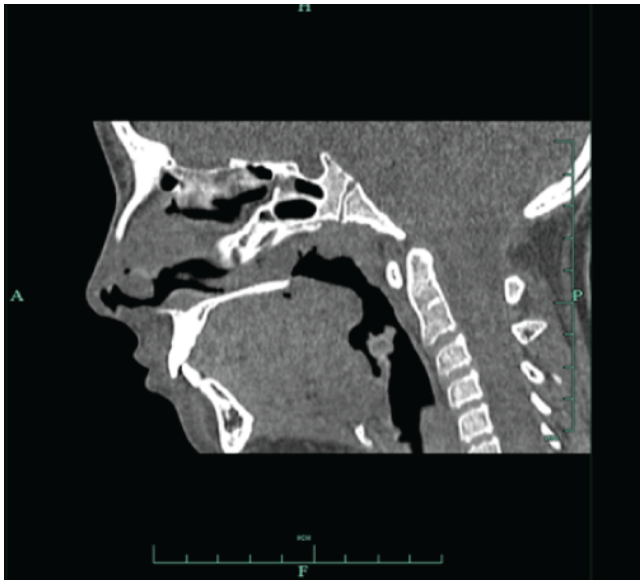

Figure 7: The computed tomography and the magnetic resonance.

Different diagnostic methods can be used to diagnose ENA. The mainstays are computed tomography and MRI also the arteriography is very helpful in getting the rightful diagnosis.This method is important in discovering the pattern of blood supply. Arteriography allows to perform selective embolization which reduce intraoperative bleeding. However computer tomography is sufficient for diagnosis. In our case the size and easy approach allowed us to remove the lesion completely without performing the arteriography.

A differential diagnosis includes fibrosedantrochoanal and ehtmoidalpolyps tumors such as meningoencephalocele, inverted papilloma, capillary hemangioma, hemangiopericytoma and solitary firous tumor [1].

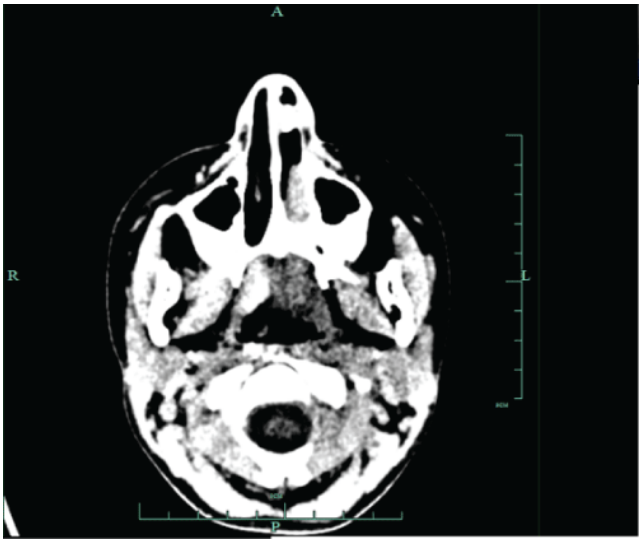

Figure 8: Pathological area of nasal septum.

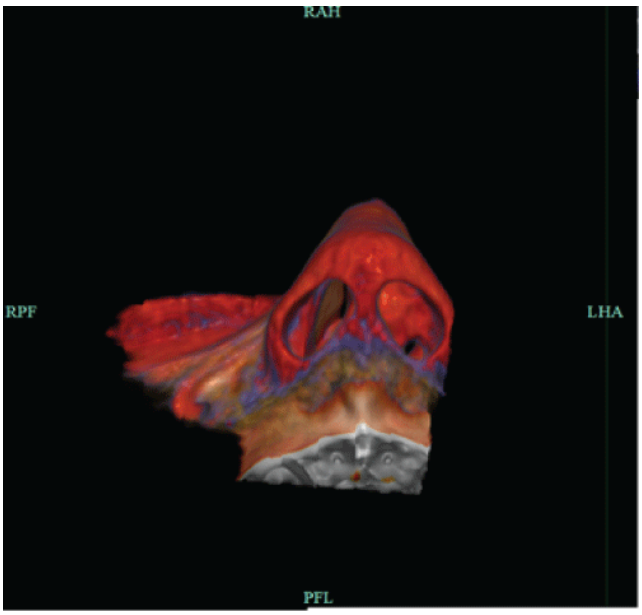

Figure 9: Nasal septum on its upper edge.

Conclusion

Angiofibroma in young female population is extremely rare. Only about 15 similar cases were found. According to literature and our own clinical experience the clinicist must be aware of the possibility of the occurrence of angiofibroma in uncommon localization in prepuberal age.

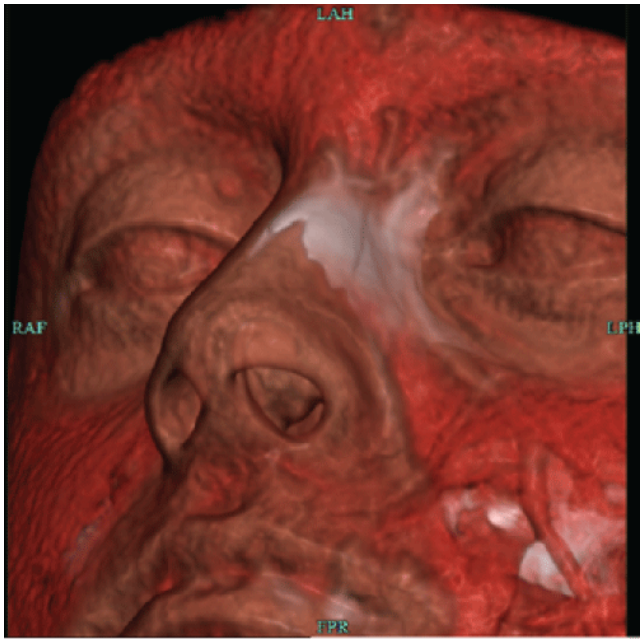

Figure 10: Partially blocked left nasal duct.

Consent Statement

Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

The authors declare that they have no competing interests.

Author's Contributions

AN has made a primary diagnosis and has removed the tumors surgically. AB has prepared the radiological evaluation of the lesion. TH has participated in its design and coordination and helped to draft the manuscript. GN is a chief of the Otolaryngology ward. All authors read and approved the final manuscript.

There are not any non-financial competing interests (political, personal, religious, ideological, academic, intellectual, commercial or any other) to declare in relation to this manuscript.

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