Clinical Characteristics of Recurrent Juvenile Nasopharyngeal Angiofibroma (JNA) After Radiation Therapy

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Abstract Juvenile nasopharyngeal angiofibroma (JNA) is a benign vascular tumor occurring in young males. Treatment of JNA is a complex procedure, especially in advanced cases with a high risk of major complications. We report the clinical, radiological and histological specialties of a rare case of recurrent JNA 3 years after radiation therapy in an 18-year-old boy. The patient underwent endoscopy assisted partial midfacial degloving approach. This treatment has been shown to be effective in advanced JNAs with minor morbidity.

Keywords Recurrent juvenile nasopharyngeal angiofibroma · Radiation therapy · Midfacial degloving

Introduction

Juvenile angiofibroma (JNA) is a rare, benign, nonencapsulated highly vascularized benign tumor that generally located in nasopharyngeal region which primarily affects adolescent males. JNA accounts for 0.5 % of all head and neck tumors and has a 1:150,000 general occurrence [1]. Nasal obstruction and epistaxis are the main symptoms of

JNA, whereas symptoms due to the involvement of adjacent structures may be present in advanced diseases. Although JNA is a benign tumor, local destructive pattern with bone erosion is a potential progress of the tumor which can result in mortal complications such as severe epistaxis, facial swelling, orbital involvement or cranial structures' dysfunction [2].

There are various treatment options but surgery is accepted as the major treatment option of JNA. Endoscopic, craniofacial and combined approaches can be used according to the tumor size, location and extent. Most of JNA can be managed by transnasal endoscopic resection with decreased morbidity, especially in the early stages of the disease [3]. Usually preoperative embolization of the major feeding vessel of tumor is done before surgery. Nevertheless, surgical management can be very difficult or morbid when there is intracranial involvement. Radiotherapy is an alternative treatment modality of JNAs.

Case Report

An 18-year-old boy was admitted to the Otorhinolaryngology Department of our hospital. His main complaints were recurrent epistaxis and nasal obstruction which resulted in snoring and sleep apnea. Three years earlier, he had undergone radiation therapy for juvenile angiofibroma. The post radiation period was partially good in the early stages after radiation therapy. His complaints have increased by time.

In the examination, it has been observed that he has severe nasal obstruction which makes it difficult to breathe especially during sleep. He has also recurrent minor epistaxis approximately every 2 or 3 days, which did not cause any changes in the hemoglobin levels. Smooth bordered

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blue–purple colored mass was seen in the endoscopic examination which totally obstructs the air way in both nasal cavities. The mass was affecting the nasopharyngeal wall, the anterior wall and the great wing of sphenoid bone and was coming into contact with the ipsilateral cavernous sinus and optic nerve and especially on the right side of the pterygopalatine and the infratemporal fossae which is staged as IIIb following Radkowski's staging system in magnetic resonance imaging (MRI) [4]. Surgical resection with preoperative embolization was planned (Figs. 1, 2).

On the operation day, the patient was intubated in the Department of Neuroradiology, where selective angiography was performed by both femoral artery punctures. The feeding main artery was selectively catheterized and embolized with using polyvinyl alcohol (Fig. 3).

The patient was transferred to the operation room for surgery. For surgery, endoscopy assisted partial midfacial degloving approach was planned. Gingivobuccal incision starting 2 cm on the left side of the midline to 8 cm right side of the midline was made. With passing submucosal tissue and perioste maxilla anterior wall of the right side of the face was exposed with preserving infraorbital nerve in the upper margin. Maxilla anterior, medial, posterior wall and septum posterior parts were removed for exposing lateral borders of mass. During exposure the open technique was used as possible as minimal bone work. Endoscopic examination was made after and especially in the upper lateral part mass was very close, attached in some parts, to the dura mater, cavernous sinus and optic nerve. The mass was totally removed without dura mater, cavernous sinus or optic nerve injury. Minimal perioperative

Fig. 1 MRI scanning of recurrent JNA

Fig. 2 CT scanning of recurrent JNA

Fig. 3 Vascularity of JNA in angiography

bleeding was controlled with cauterization, Floseal (Baxter, US) and Surgicel (Johnson and Johnson Co., US) and without any blood replacement. The cavity was filled with package. The patient's follow-up was done in the intensive care unit on the operation day and there were not any neurologic complications both related to surgery and the embolization.

There was a transient major bleeding on the postoperative first day approximately 200 cm³. Bleeding was stopped spontaneously with general measures for bleeding, such as raising the head of the patient, cold application and

resting. After bleeding package removing was postponed. The packages were totally removed on the postoperative fourth day without any complication in the operation room, instead of removing them on the postoperative second day. Nasal cavity was controlled with endoscopic examination for possible hemorrhagic points, although no hemorrhagic focus was identified. There was minor epistaxis after package removal, but no major bleeding was observed. Nasal cavity was checked on the postoperative seventh day and no hemorrhagic focus was identified. Nasal wash and lubricants were started after the second check of the nasal cavity.

The post-operative period was uneventful after the 7th day and the patient was discharged on the 15th day, showing no complications. There were no complications and signs of recurrence in the early postoperative period. Pathology result reported as recurrent juvenile nasopharyngeal angiofibroma (Figs. 4, 5).

Discussion

JNA is accepted to originate in the region above the sphenopalatine foramen at the junction between the sphenoid process of the palatine bone and the pterygoid process of the sphenoid bone. JNAs extend to the medial region such as nasopharynx, posterior ethmoid, but it is uncommon for them to extend to the lateral region such as infratemporal fossa through pterygopalatine fossa and pterygo-maxillary fissure. The treatment options consist of surgery, radiotherapy, electrocoagulation, estrogen therapy, embolization, injection of sclerosing agents, and cryotherapy [5].

Surgery is accepted as the major treatment option of JNA. Nasopharyngeal angiofibromas are relatively difficult surgical lesions. The deep-seated location, wide extensions

Fig. 4 Tumor composed of vessels and embolisation material inside

Fig. 5 Long term effect of radiation treatment as secondary fibrosis and vascular layer changes

and intense vascularity of these lesions result in difficulty in resection. Endoscopic, craniofacial and combined approaches can be used according to the tumor size, location and extent [2]. Most of JNA can be managed by transnasal endoscopic resection with decreased morbidity, especially in the early stages of the disease. Usually preoperative embolization of major feeding vessel of tumor is done before surgery.

Nevertheless, surgical management can be very difficult or morbid when there is intracranial involvement. Intracranial extension must be adequately analyzed as in most instances the tumor is extradural and extracranial, but pushes the basal dura and bone superiorly by the expansive pattern of growth. The basal bone may be eroded and thinned out by pressure [6]. Intracranial extension of JNAs are reported in different ratios, which vary from 6 to 37.5 % [7]. In advanced disease, craniofacial exposures are preferred rather than endoscopic resection, although combined approaches are used in 1/3 of the cases [2]. Craniofacial exposure includes, decreasing frequency; craniotomy, infratemporal fossa approach, lateral rhinotomy, facial degloving, Le Fort 1 osteotomy, and transpalatal approaches. However, these surgical approaches may affect facial growth due to extensive soft tissue dissection and multiple osteotomies. It may decrease the morbidity to perform endoscopy in conjunction with a limited open technique including endoscopic assisted antral window approach and Caldwell–Luc or Denker's approach [8].

Endoscopic resection should be preferred to extremely limited type IIIa or IIIb lesions and extended transfacial approach, or a craniofacial resection should be preferred for stage IV lesions [9]. Tumor involvement of the roof of the infratemporal fossa and the cavernous sinus can be difficult to access with the endoscope, leading to incomplete resection of the tumor [10]. The inability to

manipulate instruments within a confined space and of the endoscope to negotiate beyond a noncompressible tumor may be circumvented by combining endoscopic techniques with limited open approaches.

Few patients have endoscopic or endoscopic-assisted resection, it should not be concluded that these techniques are unsuitable for advanced tumors. Limits of endoscopic resection of JNAs have not been achieved, and that with further refinement of surgical techniques and instrumentation, most tumors will be amenable to endoscopic removal [3].

Surgical management of JNA with intracranial extension is complex and thus requires an expert multidisciplinary team. Although craniofacial approaches appear to be the current standard of treatment, there is increased experienced-based evidence that endoscopic resection of a large or an extended tumor is feasible in expert hands [3]. Preoperative embolization reduces intraoperative blood loss and should be undertaken in all suitable patients [11]. Subtotal tumor resection and postoperative radiotherapy does not appear to result in higher recurrence. The overall recurrence rate is 18 %. This is, however, at the expense of a high complication rate (46 %) and undoubtedly significant impact of quality-of-life issues [2].

The dissection plane between the basal dura and the tumor needs to be carefully defined during the dissection and tumor resection. The character of the tumor assists in developing a plane between the tumor and the basal dura even with a blunt dissection. Once the tumor is completely resected, the bleeding is controlled spontaneously and as the major feeders come into view they can be adequately dealt with. Incomplete tumor resection is more prone to bleeding disasters and also has a high rate of recurrence [6].

Facial paresthesia, headache, ophthalmoplegia, visual field defect, cavernous sinus injury, internal carotid artery thrombosis, intranasal crusting, CSF leak, chronic sinusitis, intranasal adhesions, epistaxis, nasal voice, cosmetic complications such as craniofacial defect which requires reconstruction, hypertrophic scarring, and alar rim necrosis, otitis media with effusion, wound infection, abscess, fistula and meningitis are the possible complications of surgery in advanced cases of JNAs [2, 12].

We added endoscopy to partial midfacial degloving for reducing incision. This helps us to avoid the development of any facial scars and related complications. Partial degloving is preferred for the widening of the surgical field.

Radiotherapy is an alternative treatment modality of JNAs. Recent studies using radiation therapy as the definitive treatment of advanced JNAs have demonstrated impressive local control rates, although complications such as cataracts, hypopituitarism and malignant transformation

have been reported. Newer radiation therapy techniques such as intensity modulated radiotherapy and Cyberknife (Accuray, Inc., Sunnyvale, CA) may decrease the development of these complications, although clinical experience remains limited. In advanced disease, subtotal resection followed by radiotherapy may decrease complications. JNAs require moderate doses of radiotherapy for durable control and cure, with a dose range of 35–46 Gy being used in various series. At these doses, there is risk of significant toxicities, particularly on the parotid gland, optic nerve, pituitary gland, temporal lobe and eye with long-term follow-up. This is valid in patients with extensive intracranial extension as in this series, with median gross tumor volume in excess of 170 cm³ [13, 14].

Recurrent diseases occur within the third postoperative year as in our case surgery is performed 3 years after radiation therapy. Current consensus for follow up is a minimum of 3 years with a 6-month interval MRI scans [1]. Incomplete surgery or radiation therapy may result in residual disease which can remain dormant. Watchful wait approach is an option for asymptomatic patients with residual disease.

In the histologic examination, we saw that the tumor was composed of vessels which do not contain an elastic layer in the fibroblastic stroma. Embolization materials were also seen in the vessels. Radiotherapy effect was also seen in pathologic specimen. Secondary fibrosis and vessels which have thickened media layer were the markers of the effect of radiation therapy.

This case report describes the characteristics and management of a rare but challenging disease which occurred 3 years after radiation therapy: growth of JNA after radiation therapy. Surgery after radiation therapy is difficult in all parts of the body. Fibrosis, adhesions and tissue defects are the main problems of surgery after radiation therapy. Due to these factors and the extent of the tumor; dura mater injury, optic nerve and cavernous sinus problems are the possible complications of surgery.

In our case careful management of this difficult case helped complete the removal of the tumor without any complications. First of all, preoperative embolization helped bleeding problems during surgery. Endoscopy assisted surgery allowed proper exposure and the removal of the tumor without any complications.

In advanced diseases, gross tumor resection cannot be achieved in all patients. Multiple surgeries or radiotherapy alone or after surgery can be performed according to the tumor's properties.

Conflict of interest None.

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