

CASE REPORTS

Transmandibular Excision of Symptomatic Large Parapharyngeal Rhabdomyoma - Case Presentation and Literature Review

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Abstract

Adult rhabdomyoma is a very rare slow-growing tumor arising in the head and neck region with a male predilection and a mean age of 61 at presentation. The most common head and neck sites are floor of mouth, oral cavity and parapharyngeal space. The case of a large parapharyngeal rhabdomyoma in a 61 years old male is detailed below from presenting symptoms, investigations, surgical management and histopathology result as well as follows up. In our case, a transmandibular approach for complete excision of this tumor was needed due to its considerable size (8.0x5.5x2.5cm) and because the tumor was encroaching the skull base. Although a rare tumor, rhabdomyoma should be considered among the differential diagnosis of neck neoplasia. Treatment with surgical excision is curative with no recurrences if fully excised.

Keywords: adult rhabdomyoma, parapharyngeal, transmandibular approach, skull base.

Rezumat

Articolul prezintă un caz cu o tumoră rară, care a necesitat, din cauza dimensiunilor mărite, o intervenție chirurgicală neobișnuită pentru o tumoră benignă, pentru a putea fi extirpată în totalitate. De asemenea, amintim existența unui patologii rare, care trebuie luată în considerare în diagnosticul diferențial al tumorilor din regiunea capului și a gâtului. Specific rhabdomiomul adultului este prevalența foarte rară, care se regăsește la nivelul capului și gâtului, cu o dezvoltare lentă. Se regăsește mai des la sexul masculin și cu o vârstă medie la prezentare de circa 60 de ani. Cea mai frecvență topografie la nivelul capului și gâtului este la nivelul planșeul bucal, cavității bucale și spațiului para-faringian. Este prezentat cazul unui bărbat de 61 de ani, cu o tumoră parafaringiană voluminoasă. Sunt detaliate: simptomele la prezentare, investigațiile efectuate, tratamentul chirurgical și rezultatul histopatologic, dar și urmărirea postoperatorie. În cazul pacientului nostru, un abord chirurgical transmandibular a fost necesar pentru a exciza în totalitate tumora din cauza dimensiunilor mărite ale acesteia, având la prezentare 8.0x5.5x2.5cm și a extensiei până la baza craniului. Deși este o tumoră extrem de rară, rhabdomiomul trebuie luat în considerare în diagnosticul diferențial al tumorilor din regiunea capului și a gâtului. Tratamentul este curativ, fără recurențe, numai dacă tumora este excizată în totalitate. Deoarece diagnosticul poate fi dificil pentru anatomopatolog, datorită rarității tumorii, este important să facem studii imunohistochimice care arată elementele caracteristice ale acestei tumori.

Cuvinte cheie: rhabdomiomul adultului, parafaringian, abord transmandibular, baza craniului.

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INTRODUCTION

Adult rhabdomyoma is a very rare slow-growing tumor arising in the head and neck region. It has a male predilection and a mean age of 60 at presentation. The most common head and neck sites for this tumor are floor of mouth, oral cavity and parapharyngeal space¹⁻³. Common presenting complaints of the patients are fullness around the angle of the mandible or airway obstruction if the tumor is present in the parapharyngeal space. Investigations should include FNA or biopsy and imaging of the neck. Immunohistological studies should be performed as due to its rarity, the rhabdomyoma sometimes could be difficult to diagnose on routine histology and also for confirmation of the diagnosis. Complete surgical excision is mandatory as recurrences occur in incomplete removal of the rhabdomyoma.

CASE REPORT

A 61 year old male (ex-smoker) presented to another institution with increasing shortness of breath over the last few months. His background medical history included sleep apnea, hypertension, and diabetes. On examination, there was no visible deformity on examination and no palpable neck mass. However, flexible laryngoscopy showed marked impingement of left parapharyngeal wall. A CT neck was performed. This revealed a very large (8x6x4cm) mass in the left parapharyngeal space (Figure 1, Figure 2).

A radiological guided FNA was reported as non-representative (simply skeletal muscle). The patient thus proceeded to core biopsy in the attempt to identify the type of tumor we were dealing with. Immunohistochemistry (IHC) supported a diagnosis of adult rhabdomyoma (strong positivity for desmin, actin and S-1001,4).

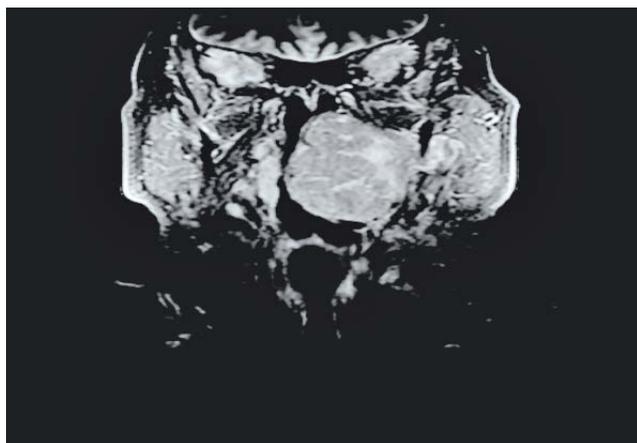


Figure 1. Sagittal MRI showing tumor compromising airway.

After multi disciplinary team (MDT) discussion, it was recommended that the patient should undergo surgical excision due to the risk of increasing airway compromise. Decision was discuss with the patient and after full explanation of the procedure and complications, patient consented for the surgical excision. This was performed by a transmandibular approach under tracheostomy cover. The tumor was found to be filling the left parapharyngeal space, extensively encroaching the skull base, and crossing the midline. Surgery proceeded uneventfully. Figure 3 is a clinical picture of the specimen *in situ* at the time of the surgery. Figure 4 shows the tumor after surgical removal.

The patient recovered well and uneventfully and the tracheostomy was removed 1 week after the operation.

Histopathology report confirmed fully excised 8.0x5.5x2.5cm adult rhabdomyoma of 84.7g.

Follow up in Out Patients department at 3 months, patient was symptomless, fully recovered without any surgical complications.

DISCUSSION

Rhabdomyoma, a rare benign tumor of skeletal muscle, is classified as cardiac or extracardiac based on its location. Extracardiac one is further divided in adult, genital and fetal type based on degree of differentiation. The adult variant of the tumor has a male preponderance (from 2:1 to 7:1 in different studies^{1,2,5}). Tumor is usually solitary but may be multifocal in up to 25% of cases^{1,6}. Symptoms depend on the site of origin. Parapharyngeal tumors may present with neck fullness around the angle of the mandible, asymmetry of the tonsil, or upper airway obstruction. Dysphagia is rare³.

Histopathological the tumor is characterized by packed, polygonal cells with abundant eosinophilic

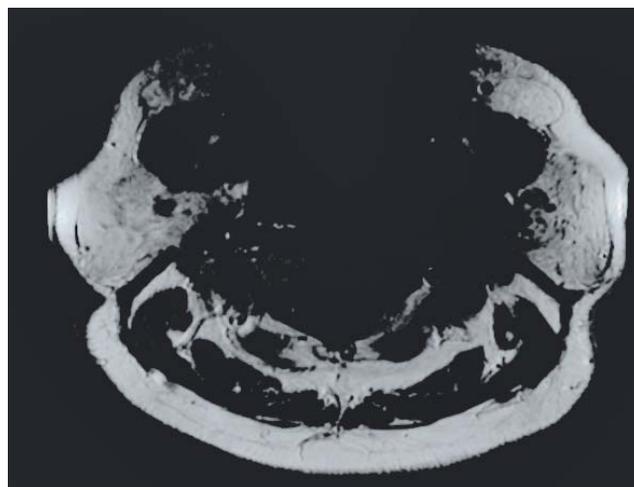


Figure 2. Coronal MRI showing tumor.

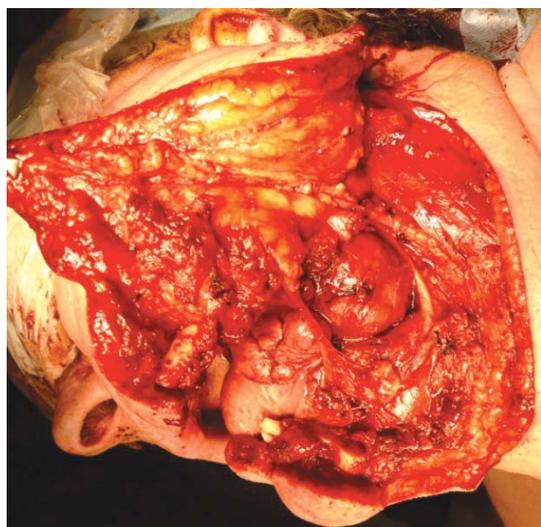


Figure 3. Tumor in situ.



Figure 4. Tumor excised.

cytoplasm and peripherally placed nuclei, peripheral vacuoles and characteristic „spider cells”¹¹.

The main histological differential diagnosis is with granular cell tumors due to the presence of granular oesinophilic cytoplasm. Other entities that should be considered are salivary gland tumors, hybernoma, acinic cell carcinoma or oncocyoma. An important tumor to be differentiated from is rhabdomyosarcoma, with a total different prognosis¹¹. The histological diagnosis should pose no problems but due to the rarity of the tumor, IHC is needed for confirmation.

Adult extracardiac rhabdomyoma is a very rare entity, with less than 50 cases described in the literature⁷. Cases as large as this one reported by us are even rarer. To our knowledge, there are only two reported cases of rhabdomyomas larger than the present case (one 8.5x2.5x1.5cm submandibular tumor described by Maglia in 2012⁸ and one 20x10 cm submental and floor of the mouth tumor reported by Bayir in 2015⁹). None of these reported large tumors needed an exten-

sive neck dissection as in a transmandibular excision or a tracheostomy.

Treatment is complete surgical excision. Recurrences are reported after incomplete resection^{1,12}.

Most benign parapharyngeal tumors can be removed through a complete transcervical approach without need for mandibulotomy or tracheostomy¹⁰. However, for very large tumors (>6 cm), a transmandibular approach is generally required for complete extirpation.

CONCLUSION

Rhabdomyoma is a very rare tumor, but should be considered among the differential diagnosis of neck neoplasia. Treatment with surgical excision is curative with no recurrences if totally excised. As diagnosis can be difficult on routine histopathology, it is important to perform immunohistochemistry studies which show the characteristic features.

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