BALKAN ORL-HNS

Solitary Fibrous Tumor of the Parapharyngeal Space Primary Surgically Treated with Poor Outcome: A Case Report

CASE REPORT
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ABSTRACT

Solitary fibrous tumor (SFT) is rare vascular tumor constituting approximately 1% of all angiogenic tumors, about 15%-20% of them occur in the head and neck region. Until now, only few cases of SFT in parapharyngeal space have been described in literature. The diagnosis relies on histological examination, while computed tomography and magnetic resonance imaging may provide useful preoperative information about tumor features. This article presents a case of SFT in a 55-year-old female where the disease was localized within the parapharyngeal space. Clinical manifestations were limited to the difficulty in swallowing and swelling in the area of the soft palate and neck. She underwent surgery using a transoral and transcervical approach and experienced a good postoperative recovery. Histopathological analysis confirmed the diagnosis and the tumor was graded as low-risk group for malignancy. Seven years later, she had relapse of the tumor, received radiotherapy, and died 2 months after that. This case underscores the need for a comprehensive evaluation because, despite the fact that the tumor was classified in the group of low malignant potential and the gross resection of the tumor was performed, the patient had a relapse that did not go away even after radiotherapy.

Keywords: Hemangiopericytoma, parapharyngeal space, solitary fibrous tumor

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Introduction

Solitary fibrous tumor (SFT), formely in literature called as hemangiopericytoma (HPC), is a rare vascular tumor, of pericytes of Zimmermann, which are baroreceptors on the luminal wall of vascular channels. Solitary fibrous tumor constitutes approximately 1% of all angiogenic tumors, about 15%-20% occurring in the head and neck, most often in sinonasal and oropharyngeal cavities.[1] Until now only a few cases of SFT in parapharyngeal space have been described in literature.

Solitary fibrous tumors are classified as benign, borderline, and malignant, depending on their histopathologic (mitotic activity, cellularity, and nuclear atypia) and clinical features (necrosis and tumor size).

The treatment of choice is radical excision.[2] The usefulness of adjuvant radiotherapy is not fully supported in literature, however radioterapy is considered for incomplete resection, malignant lesions, or inoperabile lesions. Long term follow-up is necessary as local recurrence or metastasis has been repoorted even after 7 years after treatment.[3]

Patient Information

Case Presentation

A 55-year-old female presented in October 2015 with 2 months history of difficulty in swallowing and 3 months history of swelling in the area of the soft palate and neck. There was

no information about hereditary diseases in the family history. In her personal history, she did not suffer from any systemic disease, had no surgical interventions, and had not taken any medications.

Patient also signed an informed consent for possible participation in a scientific study. This case report was approved by the institutional ethnics committee (00-456) and complied with the Declaration of Helsinki and good clinical practise guidelines.

Clinical Findings

Clinical examination showed parapharyngeal and paratonsillar submucosal elastic tumor mass with smooth mucosa above it on the left, size about 5-6 cm, and swelling on the neck on the left side.

Timeline from the Patient's First Examination to the Final Outcome

Diagnostic Assessment

CT scan head and neck and magnetic resonance angiography showed irregular oval formation, that proximally starts along the upper edge of nasopharynx and goes distally to left submandibular salivary gland, 8 × 4 cm in size (Figure 1A). Patient undervent digital substraction angiography (DSA) which showed richly vascularized soft tissue mass with feeding arterial blood vessels originating from ACE, blood vessel embolization was performed in preparation for the planned surgery (Figure 1B).

Therapeutic Intervention

Transoral, vertical incision of the soft palate and anterior palatine arch was used to access the tumor mass (Figure 2A), but more abundant diffuse bleeding appeared, so only part of the tumor was removed with this approach, and transcervical approach was indicated due to the size of the tumor. (Figure 2B). The transcervical approach began with ligation of the ACE artery above the bifurcation of the lingual artery. The tumor was completely removed by a transcervical approach. The patient underwent procedure without any complications.

Patohistological examination and immunohistochemistry showed: CD34 positive, CD99 positive, bcl2 positive, EMA negative tumor mass. The findings correspond to a mesenchymal tumor. Differential diagnosis in the first place—SFT (hemangiopericytoma). (Figure 3). Differential diagnosis is confirmed by STAT6 immunostain. If one classifies according to the latest classification, the patient gets a total of 3 points and belonged to the low-risk group for malignancy.

The surgeon's impression was that the tumor was removed completely, and there were no signs of rest or recurrence on control imaging radiological images. The tumor was low-grade and therefore radioresistant, where radiotherapy would not be of much importance.

Follow-Up and Outcome

During the first year, the patient was followed up every 3 months, and during the second year, follow-up was conducted every 4 months. In the first 2 years, there were no clinical signs of tumor recurrence, and the computed tomography (CT) scan performed in the first year was without signs of residual or recurrence. The last regular follow-up visit was in April 2017.

Next follow-up examination was in June 2022 when the patient was in poor general condition, presented with weakness and fatigue, and complained about recurrent epistaxis and impaired vision. The clinical examination then showed that the nasal passages on the left were completely filled with a pink soft tissue mass, and there was a soft tissue mass that deformed the left half of the soft palate and pharynx and pushed it to the center line. The patient underwent an magnetic resonance imaging of the head, which showed huge tumor mass that completely filled the nasal passages on the left, nasopharynx, left retrobulbar space, and left parapharyngeal space (Figure 4A). There were no regional metastases in the neck, and a CT of the chest and abdomen showed no pathological changes. It was decided not to biopsy the tumor because the tumor was very well vascularized. Biopsy of very well vascularized tumors leads to possible severe bleeding that is difficult to stop. The clinical and radiological characteristics indicated that it was the same tumor with probably more aggressive histology. The patient received only radiotherapy at a dose of 30 Gy in 10 fractions (Figure 4B) because of her poor general condition. Since being discharged from radiotherapy, the patient has not returned for follow-up examinations. She died in August 2022.

MAIN POINTS

- *Solitary fibrous tumor (SFT), formely in literature called as hemangiopericytoma, is a rare vascular tumor, of pericytes of Zimmermann origin.*
- *Solitary fibrous tumor constitutes approximately 1% of all angiogenic tumors, about 15%-20% occurring in the head and neck, most often in sinonasal and oropharyngeal cavities. Only a few cases of SFT in parapharyngeal space have been described in literature.*
- *The patient was a 55-year-old female presented in October 2015 with 2 months history of difficulty in swallowing and 3 months history of swelling in the area of the soft palate and neck. A computed tomography scan of the head and neck and magnetic resonance angiography were performed, after that, patient underwent digital substraction angiography and embolization was performed in preparation for the planned surgery. Gross resection was done through a transoral and transcervucal approach.*
- *If one classifies according to the latest classification, which implies that for a patient's age greater than or equal to 55, 1 point is given, for a tumor size of 5-10 cm 1 point, the number of mitotic activities (1-3) 1 point, and the absence of necrosis 0 points, a total of 3 is arrived at. The patient belonged to the low-risk group for malignancy.*
- *In June 2022, a tumor relapse was noted, despite the fact that the patient is classified in the group of low malignant potential. Radiotherapy was indicated; however, the patient died in August 2022.*

Discussion

Lessions of the parapharyngeal space account for 0.5 to 1% of all head and neck masses.[4] Parapharyngeal tumors are mainly composed of neurogenic or minor salivary gland origin. Vascularly originated tumors or malignancies have also have been described in this space.[5] Parapharyngeal space lesions are palpable as neck lumps when they are larger than 2.5 to 3 cm, so early detection is difficult.[4]

About 10% of all SFTs are situated in the head and neck region and usually present with sings of mass effect compression symptoms.[6] The first case of SFT in the head and neck region was described by

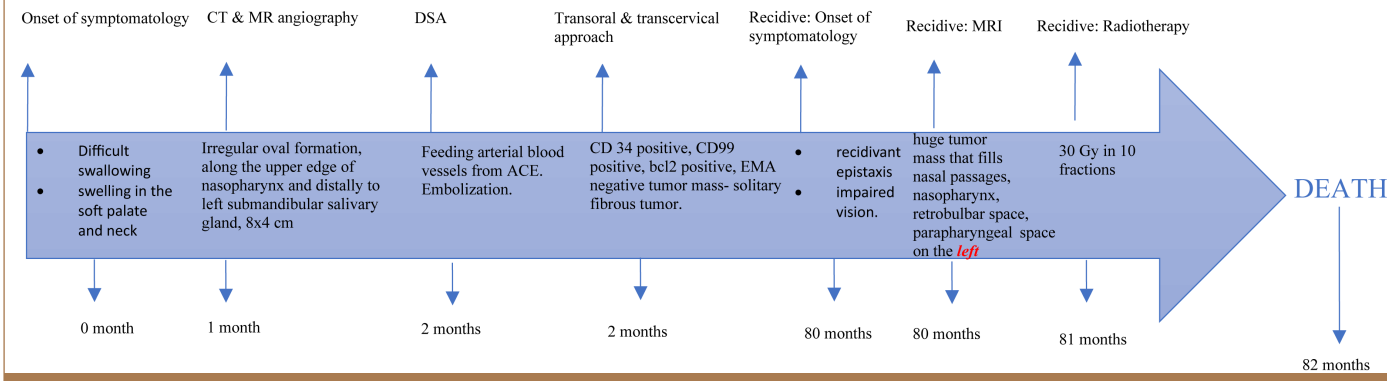

Figure 1. Computed tomography scan showing left parapharyngeal soft tissue mass (A). Digital substraction angiography showing soft tissue mass richly vascularized from ACE branches (B).

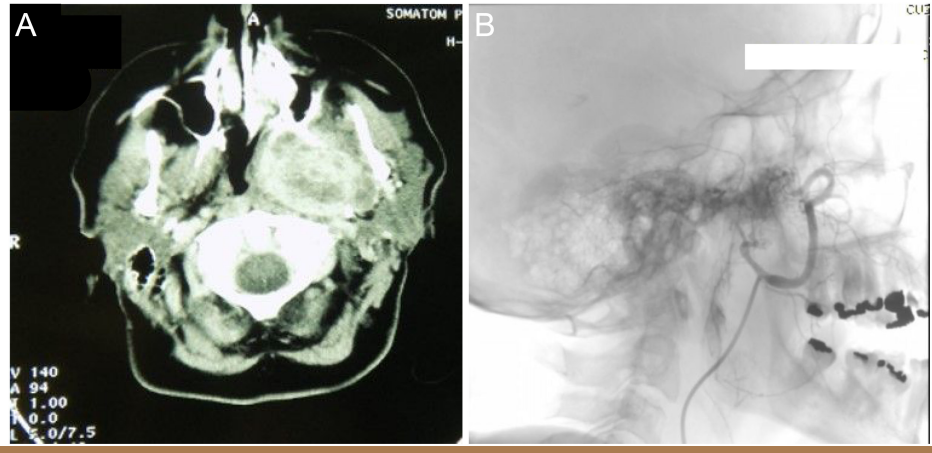

Figure 2. Transoral approach for tumor resection (A). Transcervical approach for tumor resection (B).

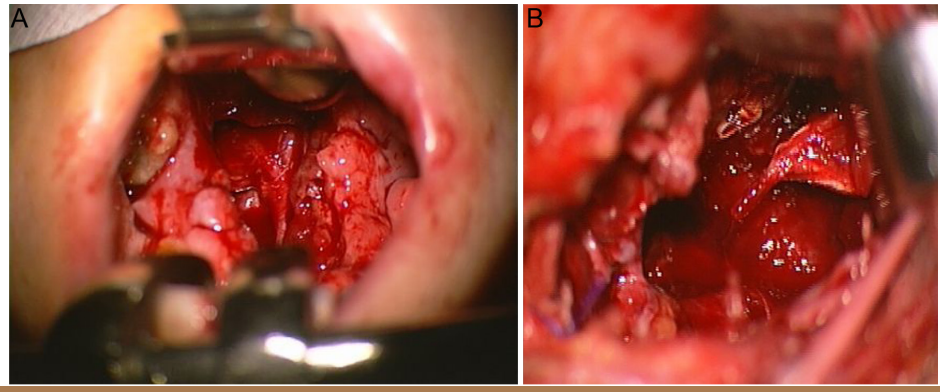

Figure 3. Microscopic view of the removed tumor—HE (×100) stain (A), immunohistochemistry staining STAT6 (×100) (B).

Witkin and Rosai in 1991.[7] Solitary fibrous tumors of parapharyngeal space are very rare, and only a few cases have been described in literature. It is frequently aggressive and has a tendency to recur and metastasize.[2] Pathologically, SFTs are characterized by abudant spindle cells and "staghorn" vascular branches.[8] Malignancy signifies the presence of necrosis, atypia, pleomorphic, or immature cells, high mitotic activity (more than 4/10 HPF), and large macroscopic mass (greater than 5 cm).[3] The clinical behavior of SFT is unpredictable and is usually described as a painless enlarging mass, but the diagnosis relies on histological examination.[9] Diagnostic imaging studies are used for the evaluation of size and infiltration of surrounding structures; magnetic resonance imaging for most precise determination of size and infiltration and CT for assessing bone infiltration. Angiography is used to confirm vascular nature of the tumor, to evaluate its vasculature, assess the risk of uncontrollable perioperative hemorrhage, and possible

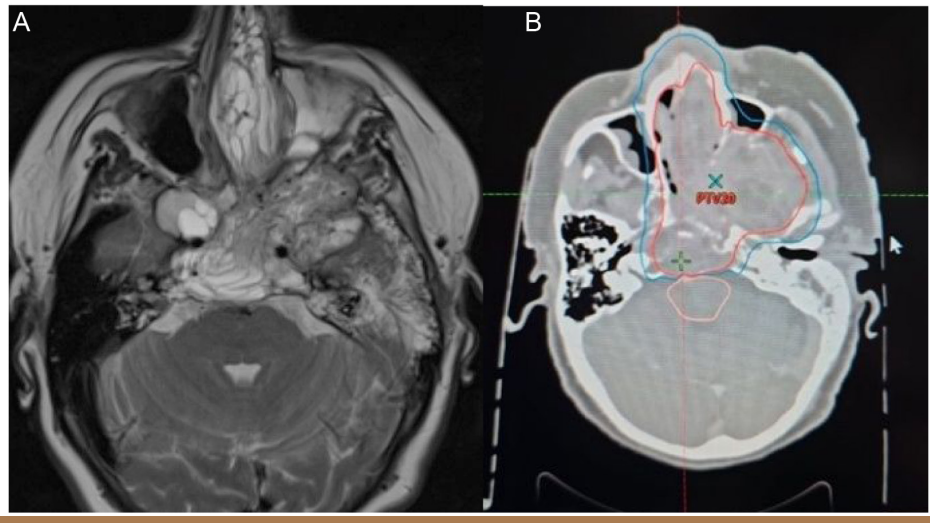

Figure 4. Magnetic resonance imaging of the head showed relapse of tumor (A). Comuted tomography of the head from radiotherapy (B).

embolization to reduce perioperative hemorrhage.[2,10] Differential diagnoses include hemangiomas, glomus tumor, histiocytomas, angiosarcoma, schwannoma, juvenile hemangioma, leiomyoma, leiomyosarcoma, mesothelioma, lipoma, liposarcoma, cystadenocarcinoma.[9,10] Immunohistochemical findings are helpful in the differential diagnosis of this disease and other tumors, and usually these tumors are CD34, STAT6, and vimentin positive, while EMA is unexpressed.[8] Radical resection of the tumor with negative tissue margins is the treatment of choice.[3,10] In the case of large, not completely resected, recurrent tumors, and potentially malignant tumors, adjuvant radiotherapy is recommended.[6] Survival rates may vary from an overall 10-year rate of 70% to a reduced 10-year survival rate in cases of >4 mitosis/10 HPF (9%), necrosis (29%), or tumor size >6.5 cm (63%).[10]

Group of authors from the Department of Radiation Oncology, The First Affiliated Hospital Zhejiang University School of Medicine, reported the twenty-third case of hemangiopericytoma of the parapharyngeal space in their paper from 2023. Seventeen out of 23 patients had a gross total resection of the tumor and 6 of them had subtotal resection. Seven of them received adjuvant radiotherapy.[11] This makes this case the twenty-fourth reported case of SFT in the literature, and the eighteenth that was treated by gross resection of the tumor. When recurrence occurred, the patient received radiotherapy at a dose of 30 Gy in 10 fractions and despite that, passed away 2 months later.

Despite the fact that the patient is classified in the group of low malignant potential and that gross resection of the tumor was performed, the patient had a relapse that did not go away even after radiotherapy. This makes this case even more special and worth considering.

Data Availability Statement: *Datasets regarding the case being discussed are available on request from the corresponding author.*

Artificial Intelligence (AI) Use Declaration: *The authors declared that no artificial intelligence (AI) tools were used in the generation, writing, or editing of this manuscript, and all content was produced solely by the authors.*

Ethics Committee Approval: *This case report was approved by the Ethics Committee of Institutional Ethics Committee (no: 00-456).*

Informed Consent: *Written informed consent was obtained from the patient who agreed to take part in the study.*

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