Management of head and neck tumors in patients with Haemophilia A. A Case report

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Αντιμετώπιση όγκων κεφαλής και τραχήλου σε ασθενείς με αιμοφιλία Α. Αναφορά περίπτωσης

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Case Report
Αναφορά Περιστατικού

SUMMARY: In this article, we report a case of a 61 year old male with hemophilia A, who was presented in our clinic with two simultaneous sizable tumors. A Warthin tumor in the left parotid gland and a sebaceous cyst in the submental area region. The patient had these tumors for over than 20 years because he was afraid the potential bleeding intraoperatively due to his underlying hemophilia A. After clinical examination, blood test and CT scan the patient underwent partial superficial parotidectomy for Warthin tumor and extracapsular dissection of the submental sebaceous cyst. The mainstay of the patient's management was the defintion of Haemophilia A status and substitutionwith VIII factor. Also anesthetic techniques associated with bleeding were avoided and hemostasis was carried out very carefully. Thus, any severe bleeding was not presented in surgery field and the patient did not develop postoperative bleeding or facial nerve weakness symptoms. In conclusion, under specific prerequisites surgical procedures can be performed safely in patients with hemophilia A.

KEY WORDS: *Hemophilia, Warthin Tumor, Sebaceous cyst, Management, Surgical treatment*

ΠΕΡΙΛΗΨΗ: Σε αυτό το άρθρο, αναφέρουμε μια περίπτωση άνδρα 61 ετών με αιμορροφιλία Α, ο οποίος παρουσιάστηκε στην κλινική μας με δύο μεγάλους όγκους ταυτόχρονα. Ένας όγκος Warthin στην αριστερή παρωτίδα και μια σμηγματογόνα κύστη στην περιοχή της υπογενείδιας περιοχής. Ο ασθενής είχε αυτούς τους όγκους για περισσότερα από 20 χρόνια επειδή φοβόταν την πιθανή διεγχειρητική αιμορραγία λόγω της υποκείμενης αιμορροφιλίας Α. Μετά από κλινική εξέταση, εξέταση αίματος και αξονική τομογραφία ο ασθενής υποβλήθηκε σε μερική επιπολής παρωτιδεκτομή για τον όγκο Warthin και εξωκαψική εκτομή της υπογενείδιας σμηγματογόνου κύστης. Ο βασικός άξονας της διαχείρισης του ασθενούς ήταν ο ορισμός της κατάστασης Αιμορροφιλίας Α και η υποκατάσταση με τον παράγοντα VIII. Επίσης, οι τοπικές αναισθητικές τεχνικές που σχετίζονται με τοπική αιμορραγία αποφεύχθηκαν και η αιμόσταση έγινε πολύ προσεκτικά. Έτσι, σοβαρή διεγχειρητική αιμορραγία δεν παρουσιάστηκε και ο ασθενής δεν εμφάνισε μετεγχειρητική αιμορραγία ή συμπτώματα αδυναμίας λόγω κάκωσης του προσωπικού νεύρου. Συμπερασματικά, κάτω από συγκεκριμένες προϋποθέσεις οι χειρουργικές επεμβάσεις μπορούν να πραγματοποιηθούν με ασφάλεια σε ασθενείς με αιμορροφιλία Α.

ΛΕΞΕΙΣ ΚΛΕΙΔΙΑ: *Αιμορροφιλία, Όγκος Warthin, Σμηγματογόνος κύστη, Διαχείριση, Χειρουργική θεραπεία*

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INTRODUCTION

In this article, we report a case of a 61 year old male with hemophilia A, who was referred to our clinic with two simultaneous sizable tumors a Warthin's tumor of the left parotid gland and a large sebaceous cyst in the submental region.

Hemophilia A is a clotting disorder associated with bleeding due to factor VIII deficiency. The clinical presentation appears with a significant fluctuation in accordance to FVIII presence and activity [1,2].

Warthin's tumor is the second most common salivary gland tumor after pleomorphic adenoma [3,4,5]. Surgical treatment remains the gold standard therapy for Warthin's tumor and has two options: extracapsular dissection and partial superficial parotidectomy [4]. Sebaceous cyst, also known as epidermoid cyst, is a benign encapsulated, subepidermal nodule filled with keratin material. The mainstay therapy is complete surgical excision in order to remove the cyst wall intact. [6].

CASE PRESENTATION

A 61-year-old Caucasian man was referred to the outpatient department of Oral and Maxillofacial Surgery of "Evagelismos" Hospital for an enlarging mass over the left parotid gland and an enlarging mass in the chin region. The patient had multiple comorbidities including hemophilia A, type II diabetes, atrial fibrillation and hypertension.

The patient had these tumors for more than 20 years because he was afraid of the intraoperative bleeding due to hemophilia A. He was systemically well with no weight loss, salivary pain, odynophagia, dysphagia, xerostomia, hemoptysis or facial weakness. Physical examination demonstrated that the masses were superficial, soft, painless and mobile. The skin overlying the surface of the masses was normal with no discoloration or tethering and the skin temperature was not elevated. There was no cervical lymphadenopathy and the facial nerve was intact (Figure 1).

A CT scan described a tumor 5x6x8cm with cystic elements in the left parotid's tail, which extended inferior in the submandibular region and the neck, as well as a large cyst measuring 8cm in the right submental region (Figure 2). The diagnosis was further supported by the results of fine-needle aspiration biopsy, which showed features suggestive of a Warthin's tumor in the parotid gland and a sebaceous cyst in the submental region.

As reported previously the patient of this case report suffered from hemophilia A, which affects blood clotting in conjunction with the level of absence of blood clotting Factor VIII. His blood test results were as followed prior to surgery and immediately pre op following prophylactic treatment (Table 1). In coordination with the hematologic unit the patient received 3000IU FVIII (HAEMOCTIN®) for prophylaxis 30 minutes before surgery.

Afterwards, the therapeutic schedule was determined as 1000 IU every 12 hours for the first 2 postoperative days and 1000IU every 24 hours until the 8th postoperative day, in order to avoid intraoperative and postoperative bleeding.

Intraoperatively prior to skin infiltration of local anesthetics with a vasoconstrictor (epinephrine 1:100000) was injected subcutaneously to aid in hemostasis. The incision for the chin mass was placed in a natural skin crease of the neck. The tumor was beneath the skin and was revealed right away after the skin incision underneath the subcutaneous tissue. The overlying tissue was carefully dissected and the mass was completely removed (Figure 3 A-C).

The incision for the parotid tumor was placed in a preauricular crease and passed inferiorly, crossing the base of the tragus and passing posteriorly behind the lobe of

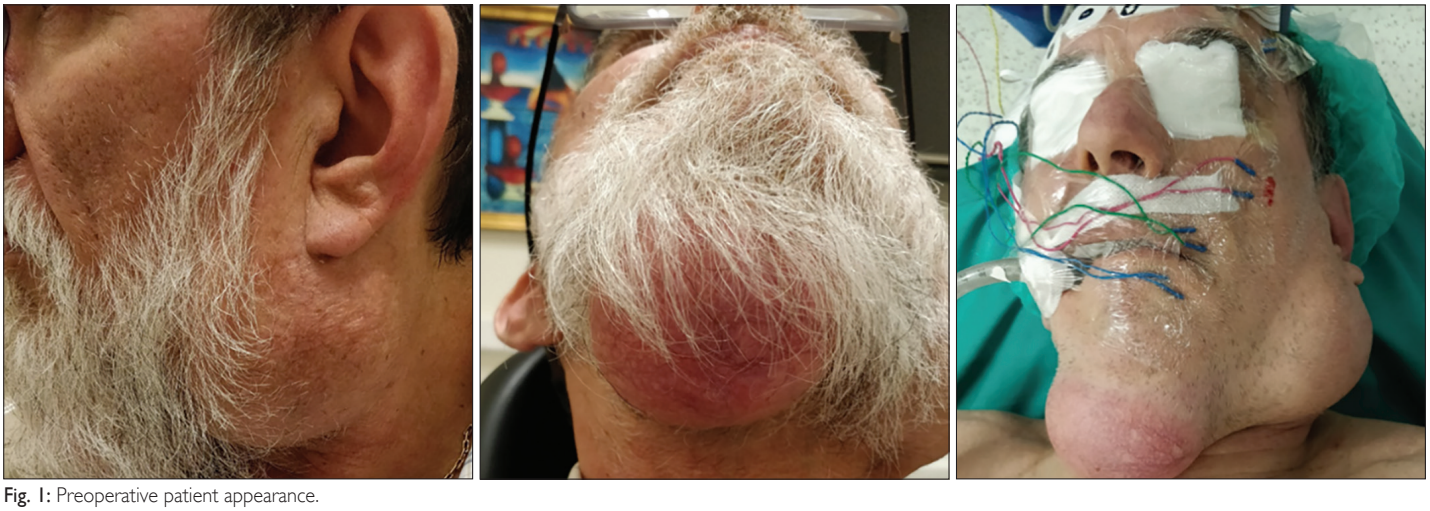

Fig. 1: Preoperative patient appearance.

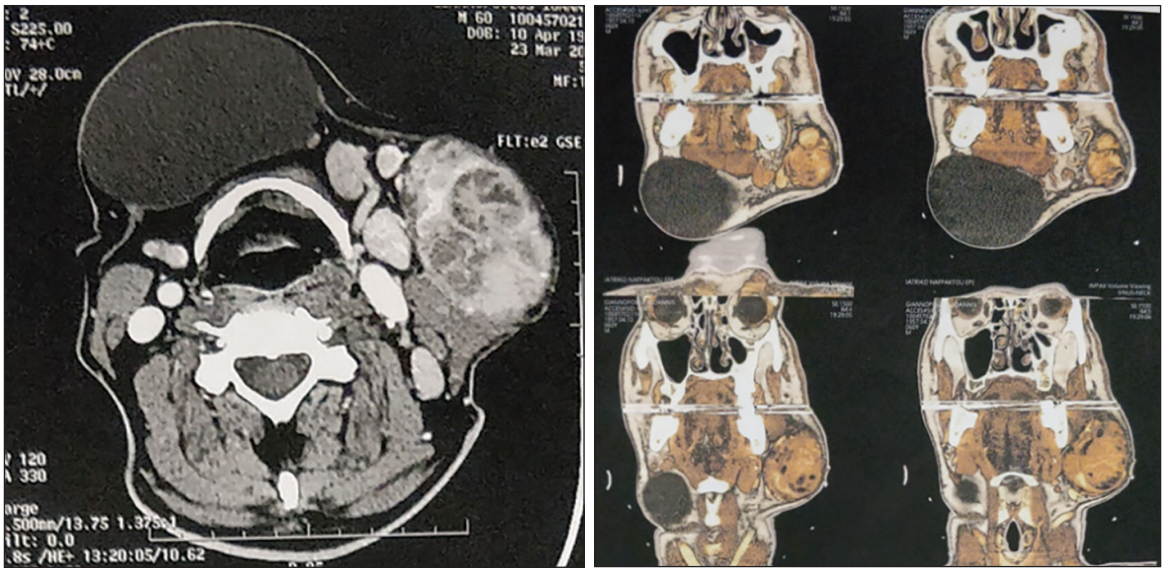

Fig. 2: Preoperative imaging evaluation. CT scan showing the submental mass and the tumor in left parotid gland.

the ear. Then it extended inferiorly from the mastoid to continue in a natural skin crease of the neck. The skin flap was raised in the plane of the pre-parotid fascia in the preauricular region and in the subplatysmal plane in the neck and the mass was revealed. The facial nerve was identified between the pointer and the mastoid process, about 1cm deep; then the overlying tissue was carefully dissected, as the tumor was attached in the upper branch. After surgical removal of the tumor, parotid tissue and fascia were reapproximated and sutured to the anterior border of the sternocleidomastoid muscle (Figure 4 A-C). All bleeding points had been meticulously controlled with diathermy while larger vessels were ligated.Two vacuum drains were inserted under the flaps and the wounds carefully closed in layers. A firm pressure dressing was placed to prevent any collection of blood or saliva under the flap. The patient didn't present postoperative facial nerve weakness symptoms.

The pressure dressing was removed at about 48 hours and the vacuum drains at 72 hours. Skin sutures were removed after 7 days (Figure 5).

The postoperative pathological diagnosis was Warthin's tumor in the parotid region and sebaceous cyst in the chin region.

DISCUSSION

Hemophilia A is a clotting disorder associated with bleeding due to factor VIII deficiency. It may be X linked recessive inherited or acquired, which can emerge from spontaneous mutations. It has an estimated frequency

Table 1: Patient's blood test results

	Pre Op	Immediately Pre Op After FVIII
PT (%) NR 90-110	37	76
aPTT NR 29-40	72.2	39
FVIII NR 65-150	20	–
FVIII Ag NR 70-150	8	–

Patient's bloodtest results related to HaemophiliaA screening. PT: Prothrombin Time, aPTT: Activated Partial Thromboplastin Time, FVIII: Human coagulation factor VIII, FVIII Ag: Human coagulation factor VIII related antigen, NR: Normal Range.

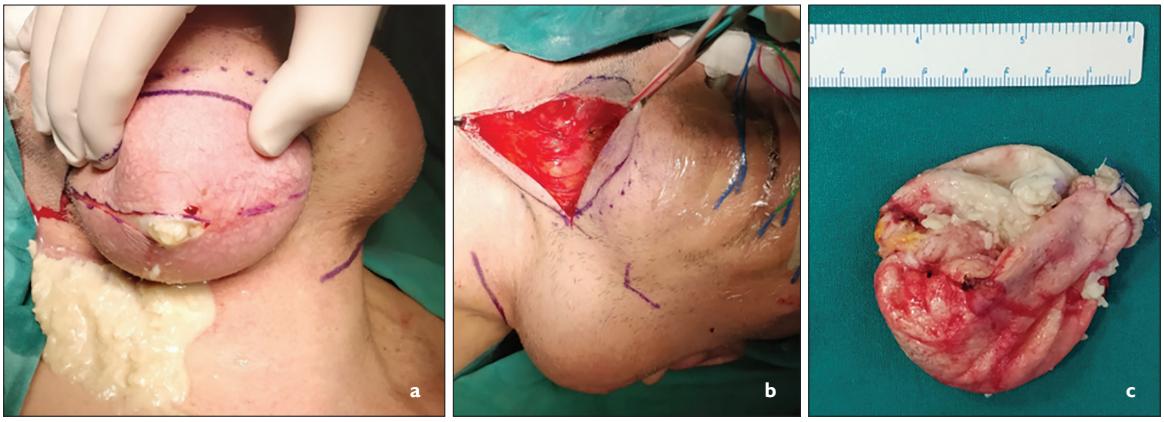

Fig. 3: a: Intra-operative image showing the blowout of the content of sebaceous cyst after the initial incision. b: The surgical field after the excision of submental mass c: Macroscopic appearance of the surgical specimen.

of 1 in 10000 births, with male predominance and accounts for 85% of the total hemophilia population [1,2]. The severity of the disorder depends on the FVIII activity. In order to measure this there are three different methods, either measure the FVIII Ag in plasma, or with chromogenic assay or with I stage clotting assay [7]. Subsequently, the severity is categorized accordingly, normal FVIII activity from 50 to 100%, mild from 5 to 40%, moderate from 1 to 5 % and severe less than 1% FVIII activity [2]. Consequently, the patient in our case report was suffered by mild hemophilia A. Moreover, bleeding time and prothrombin time (PT) are expected to be within normal range, while activated partial thromboplastin time (aPTT) is prolonged [2].

The clinical presentation may vary according to the severity, from spontaneous bleeding to clinically silent or after trauma or surgery [2]. Successful management for patients, who are scheduled for elective surgery, involves collaboration between the surgeon and the hematologic unit for prophylactic replacement therapy to be administered, which may involve desmopressin (DDAVP), FVIII plasma concentrates and cryoprecipitate. Antifibrinolytic drugs are used to control mucocutaneous bleeds or after dental extractions [2]. In our case, the patient received the prophylactic dose of VIII factor. Anesthetic techniques associated with bleeding (naso- tracheal intubation, central lines) were avoided and hemostasis carried out very carefully with bipolar diathermy. As a result we didn't have any severe bleeding in surgery field and the patient didn't present any problems during the postoperative period.

Warthin's tumor accounting for up to 17% of all salivary gland tumors [8,9]. In the majority of cases it is located at the inferior pole of the parotid gland, but also it presents multicentric occurrence [4,5]. The risk of malignant transformation for this type of tumor is less than 1% [5]. Clinically, the tumor does not grow aggressively and the first clinical manifestation is a swelling of the parotid region. Investigation of a suspected Warthin's tumor commonly involves MRI scan and fine needle aspiration (FNA) biopsy [3, 5]. The gold standard for Warthin's tumor treatment is surgical excision [3,4]. The extent of the operation for Warthin tumor remains a controversy between extracapsular dissection and partial superficial parotidectomy [4]. The most common and most feared complication during superficial parotidectomy is the fa-

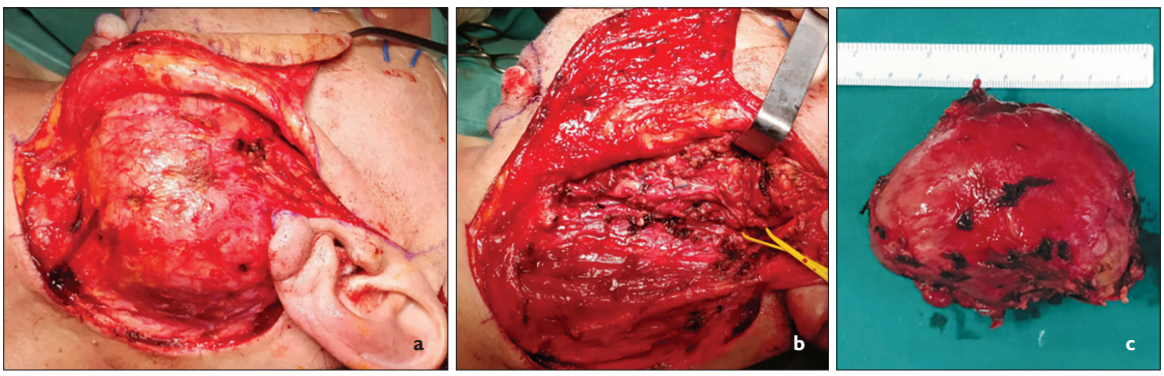

Fig. 4: a: Intra – operative image showing the reveal of parotid mass b: The intact facial nerve after mass excision c: Macroscopic appearance of the surgical specimen.

Fig. 5: Post – operative image showing the clinical aspect after tumors removal with facial nerve preservation.

cial nerve injury. In our case the tumor was removed with superficial parotidectomy and the patient didn't present facial nerve weakness.

Sebaceous cysts are benign lesions of the skin. These lesions may grow anywhere on the body; about 7% appear in the head and neck [6,10]. Approximately 1% of these cysts have been noted to have a malignant transformation to squamous cell carcinoma (SCC) or basal cell carcinoma (BCC) [6, 10]. Ordinarily, the physical examination reveals a non-fluctuant, compressible mass with a central dark opening (punctum). Generally, they are asymptomatic; but if they rupture clinical signs will be observed, such tenderness to palpation, erythema, swelling, pain and a foul-smelling yellowish cheese-like material discharged from the skin. In the evaluation of epidermoid cysts laboratory examination and radiographic tests are not necessary. The main step for diagnosis is the clinical examination combined with a good history [6, 11]. The surgical excision is the only treatment and the completely remove is very essential to prevent recurrence [11]. In our case, the incision was made in a skin tension line in submental region. Although the cyst was removed after carefully blunt dissection we cannot avoid the cyst rupture due to their large dimensions. Nevertheless, the cyst wall was completely removed.

CONCLUSION

On the whole, patients with hemophilia can be scheduled for elective surgery after cautious preparation. A thorough medical history, clinical examination, blood exams and also collaboration between surgeons and hematologists are necessary to avoid complications. Furthermore, intraoperative meticulous hemostasis is crucial and postoperative follow up is necessary from both medical teams. Under these prerequisites, surgical procedures can be performed with safe in patients with hemophilia.

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