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ORIGINAL ARTICLE

Management of vascular malformations of the parotid area

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KEYWORDS
Parotid tumor;
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Summary
Objectives: To describe our experience in the management of vascular malformations of the parotid area.
Materials and methods: This was a retrospective study. Among 614 parotidectomy performed between 1998 and 2008 at our institution, 10 cases (1.6%) of vascular malformations have been identified. Clinical features and management of these patients were analyzed.
Results: Clinical presentation was usually related to that of a benign, slow-growing and asymptomatic tumor. There was a marked female predominance (90%). In any case, the diagnosis of vascular malformation could be obtained with certainty preoperatively. Surgical excision was performed most often referred to diagnosis. Sixty percent of vascular malformations were located in the superficial lobe of the parotid gland. On the histological we found a classic look with benign vascular proliferation of endothelial cells in the walls. The vessel lumen was either the head of a congestion or thrombosis or calcification (phleboliths).
Conclusion: Vascular malformations of the parotid gland, rare disease, are mainly venous. The terminology is based on clinical data, scalable, histological and hemodynamic as classified by the International Society of Study of Vascular Anomaly (ISSVA). Despite advances in imaging including MRI they remain difficult to diagnose. The treatment of reference is surgical excision.

Introduction

As the 21st century begins, vascular anomalies (localized defective angiogenesis) remain one of the least well-understood entities encountered in clinical practice. The reason is simple: there has been no animal model for studying the underlying pathological mechanisms and developing therapeutic models. Moreover, the use of confusing terminology has led to insufficient diagnostic techniques, inadequately adapted treatments and poorly oriented research. The generalized term 'angioma' is still used to describe both tumors and vascular malformations. The international classification resulting from the 10th Workshop of the International Society for the Study of Vascular Anomalies

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(ISSVA), held in Rome in 1996, was based on clinical, evolutive, histological and hemodynamic elements, and should now be used [1]. It describes two large categories of tumors and vascular malformations: 'immature' tumors, such as hemangiomas, which are observed in infants and, after a period of growth, eventually regress spontaneously; and 'mature' tumors, which do not regress, but continue to develop throughout life. Vascular anomalies are classified according to the type of vessel involved — hence, capillary, venous, lymphatic and arteriovenous malformations. Combined forms are also found, including venolymphatic and arteriovenolymphatic malformations. These malformations are the focus of the present work. Vascular malformations of the parotid gland are extremely rare, and constitute a distinct entity of parotid pathology that requires specific diagnostic tools and management. Around 50 cases (mainly venous malformations) have been described in the literature so far [2], mostly as individual case reports.

The purpose of the present study was to review our experience over the past 10 years to clarify the diagnosis and management of these vascular malformations.

Material and methods

The medical records of all adult patients who underwent parotidectomy at our institution between 1998 and 2008 were reviewed. During this 10-year period, of the 614 parotidectomy procedures performed, 10 cases of vascular malformation were identified. Among these 10 cases, there were nine cases of venous malformation and one of arteriovenous malformation.

Data collected were age, gender, functional and clinical signs, diagnostic procedure, histological features and patient outcome. Descriptive statistical analyses were also performed.

Results

Of the 614 patients, 462 (75%) had benign tumors and 152 (25%) had malignant tumors. Vascular malformations represented 1.6% of all parotid tumors and 2.1% of all benign tumors in our series.

Age and gender

Our cohort of 10 patients presented a median age of 42 years (range: 19—54 years). Female gender predominated (nine women and one man). Although no predisposition (genetic, hormone profile) was found, this was not systematically searched for at history-taking. Similarly, there were no cases of associated extraparotid vascular malformations.

Tumor localization

There was no side predominance (six on the right and four on the left) and no bilateral cases. Six of the 10 cases (60%) were, situated in the superficial lobe of the parotid gland, two (20%) were in the deep lobe with parapharyngeal extension and two (20%) were within the masseter muscle.

Clinical presentation and diagnostic approach

Our patients attended our clinic for various reasons. Five patients (50%) reported the fortuitous discovery of an asymptomatic mass on self-palpation that, in one case, was following surgery for a contralateral cholesteatoma. The other five patients (50%) complained of a functional problem: pain ($n = 2$); dyspnea ($n = 1$); fistula to the skin ($n = 1$); and dysphagia with lateral expansion of the velum ($n = 1$).

The clinical presentation was generally suggestive of a benign tumor: slow growth; soft consistency; mobile; no preoperative facial palsy; no infiltration of the skin; and no enlarged neck nodes on palpation. Nevertheless, one patient (10%) had a hard tumor that had infiltrated the skin and soft tissues, and fistulized to the skin. Magnetic resonance imaging (MRI) was performed in all cases preoperatively, but the initial diagnosis was established by ultrasound (US) examination in two patients and on the basis of the computed tomography (CT) scan with contrast injection in three patients. However, none of the imaging methods allowed a definitive diagnosis of vascular malformation preoperatively. The suggested diagnosis before surgery was a mixed tumor in three patients (30%), cystadenolymphoma in two (20%), tumor of undetermined nature in four (40%) and sarcoma or neurinoma in one (10%). MRI showed gadolinium uptake and hyperintense T2-weighted signals in all 10 patients. The contours were irregular on the T2 images for 30% of the patients. The hyperintense T2 signal correlated with a hyperintense T1-weighted signal in four patients (40%); for the six others, it was associated with a hypointense T1 signal (Fig. 1). Needle aspiration was performed in three patients and was non-contributive in all three cases.

Treatment

The diagnostic work-up was unsuccessful in establishing the diagnosis of vascular malformation preoperatively in all 10 patients in our series. Vascular malformation was nevertheless among the preoperative diagnostic hypotheses. The time from symptom onset to surgery was 38.4 months on average (range: 1—84 months), and the decision to operate based on either the clinical presentation [dyspnea ($n = 1$), pain ($n = 2$), fistulization ($n = 1$) and dysphagia ($n = 1$)] or the preoperative diagnostic work-up which was not in favor of vascular malformation.

In our series, the surgical procedure was exofacial parotidectomy in five patients (50%), total parotidectomy in four (40%) and enucleation in one (10%). Anterograde exposure and dissection of the facial nerve was performed in all patients; the facial nerve was also preserved in all cases.

Total tumor resection was achieved in nine patients (90%). The one case of partial resection was related to the excessive risk of injury to the facial nerve; treatment was successfully completed by radiological embolization late after surgery. Except for this patient, the surgical procedures were uneventful; none of the surgery reports mentioned abundant bleeding or difficult dissection of the facial nerve.

Mean hospital stay was 5 days (range: 3—17 days), with a mean follow-up of 3 years (range: 9 months to 10 years).

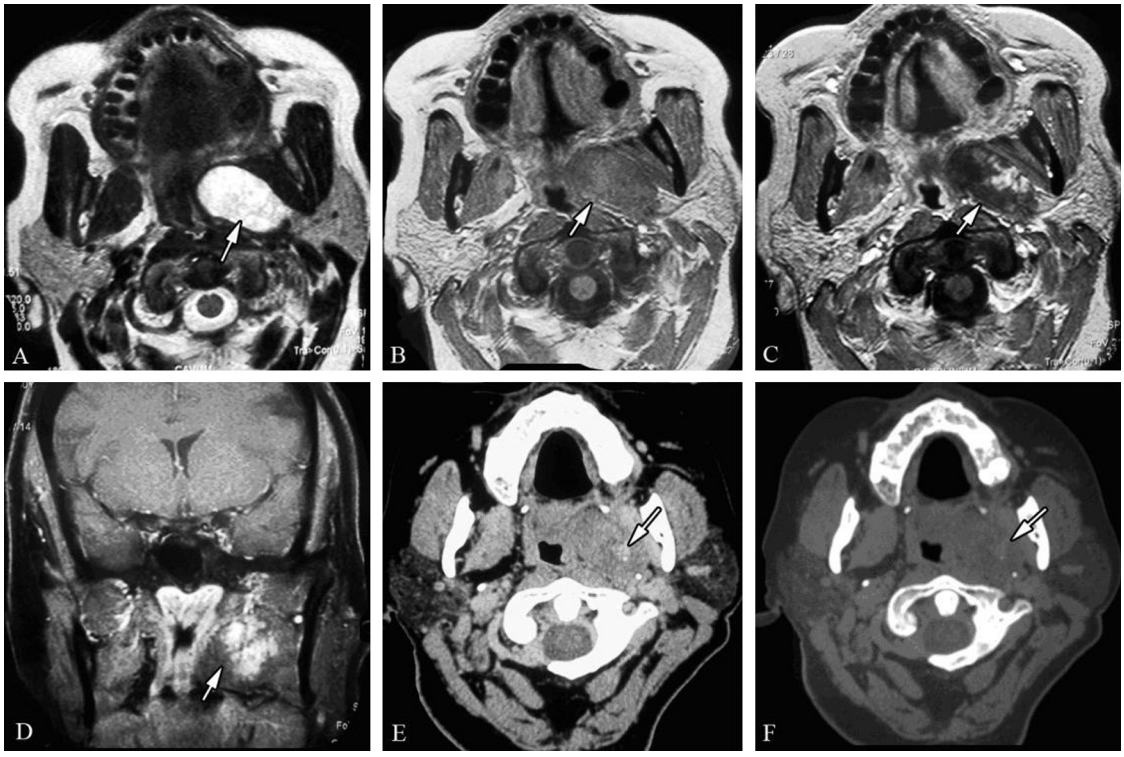

Figure 1 MRI scans: T2-weighted (A), axial T1-weighted (B), axial T1 with gadolinium (C), and coronal fat-saturated T1 with gadolinium (D); CT parenchymal window without injection (E) and bone window (F). A tissue lesion (A—F, arrows) is seen in the prestyloid parapharyngeal space located in the extended parotid space, displacing the lateral pterygoid muscle anteriorly and styloid diaphragm posteriorly. This formation produced a hyperintense T2 signal (A) and a hypointense T1 signal (B), with spotty enhancement (C, D). The formation was calcified on the CT scan (E, F).

Transient postoperative complications were noted in two patients (20%). One presented with paresis of the frontal branch and Frey syndrome, which totally resolved in 6 months. The other patient presented with facial nerve palsy affecting the mandibular branch that resolved in 3 months; this case was a repeat procedure after an initial operation performed 20 years earlier at another center. There was also one case of recurrence in yet another patient (10%). This patient had a tumor in the deep lobe that extended to the parapharyngeal area with fistulization. Recurrence was diagnosed within 3 years and required preoperative embolization followed by repeat surgery with sacrifice of the facial nerve and reconstruction using a pedicled latissimus dorsi flap. The primary tumor was very large, as was the recurrence.

Histology

Mean tumor length was 40 mm (range: 5—90 mm) and mean width was 20 mm (range: 7—50 mm). Preoperative histology was available for all 10 cases and favored a vascular malformation in six. For the four others, the diagnosis was established on examination of the surgical specimen.

The final histology report noted benign vascular proliferation composed of variably sized dystrophic vessels with more or less thick walls bordered by regular endothelial cells with no cell atypia. The vessel lumens were congested, thrombosed (Fig. 2) or sometimes calcified.

For the fistulized lesion, there was a poorly defined vascular lesion of the skin composed of variably sized vessels, often with thick walls. These vascular structures exhibited a number of anastomoses. The final diagnosis was intraparotid arteriovenous malformation (Fig. 3).

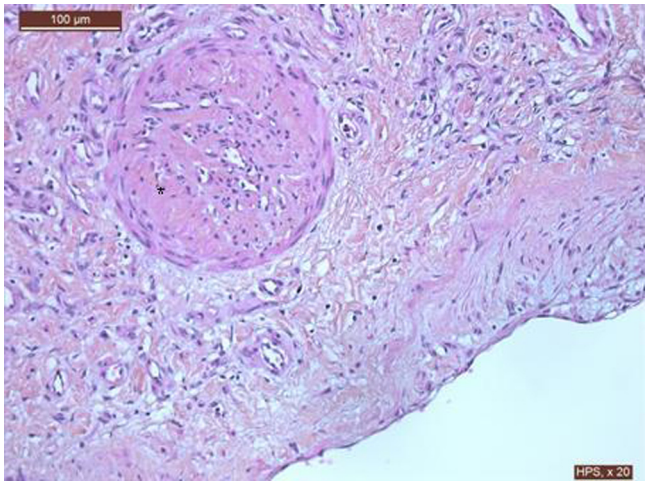

Figure 2 Vascular proliferation with muscularized vessel walls and thrombus (*) in the vessel lumen. Hematoxylin phloxine saffron stain, × 20.

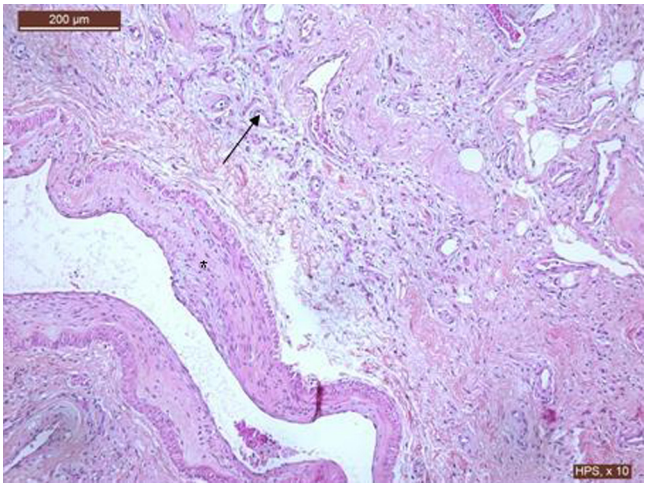

Figure 3 Arteriovenous malformation: note the thick arterial wall with an elastic limiting layer (*) and small-sized venous structures (arrow). Hematoxylin phloxine saffron stain, × 10.

Discussion

The classification of 'angiomas', vascular tumors and malformations, has been somewhat confused in the past. For this reason, the ISSVA recently adopted a classification system that distinguishes vascular tumors (lesions with clear manifestations of cell proliferation) from vascular malformations (due to innately perturbed vascular morphogenesis). These latter are classified according to the type of vessel involved — hence, capillary, venous, lymphatic and arteriovenous malformations.

In young children, hemangioma (a vascular tumor) is the leading cause of parotid tumefaction [3,4] whereas, in adults, vascular malformations affecting the parotid gland are rare. In our series of adult patients, parotid malformations accounted for only 1.6% (10 out of 614 patients) of all parotid tumefactions collected over the 10-year period from 1998 to 2008. In the literature, the frequency has been noted at 0.5% for Bears et al. [5], who reported 760 parotid tumors, and at 0.6% for Byars et al. [6], who reported 460 parotid tumors. There is a clear female predominance (nine of 10 patients in our series), but no apparent side predominance. Diagnosis is made around the fourth decade of life. All of the patients in our present series had a solitary tumor with no associated predisposing factors. The lack of an animal model makes it difficult to study these vascular anomalies. However, molecular genetics has recently enabled the isolation of genes responsible for the development of certain vascular malformations and vascular tumors (mainly hemangiomas) [7], although no genetic studies were conducted in our patients.

All areas of the parotid gland may be involved; however, in our series, 60% of the lesions were in the superficial lobe, 20% in the deep lobe and 20% within the masseter muscle. The last two cases were included in our series because they are managed in the same way as parotid tumors (and only one was diagnosed preoperatively). In the literature, the masseter muscle is the primary site of vascular malformations within the facial muscles, accounting for 4.9% of all intramuscular vascular malformations [8].

Clinically, these vascular malformations are seen as a slow-growing, soft, mobile mass, with no facial nerve palsy and no lymph-node enlargement or cutaneous infiltration. The majority of patients develop few or no symptoms. However, one of our patients had a hardened tumor that had infiltrated the skin and soft tissues, and fistulated. This was a poorly defined, non-encapsulated arteriovenous malformation. The severity of this type of vascular malformation is mainly due to its local blood-shunting effect with the risk of skin necrosis (fistulization, as observed in our patient).

Symptoms, when present, were related to either the longstanding nature of the tumor (dyspnea or dysphagia with a raised half soft palate) or to microthrombotic events (pain-causing phleboliths). These phleboliths are a consequence of venous stasis and are never the cause of clinical tumefaction [9]. Dempsey et al. [10] described a specific clinical sign, the 'turkey wattle sign', found in 50% of their patients. Tumor volume increases with the Valsalva maneuver or when the head is tilted forward. We did not look for this sign in our patients.

MRI and needle aspiration are the most useful complementary explorations when searching for malignancy. CT with contrast injection and US are first-intention explorations for the initial work-up. A specific aspect of venous malformations is the presence of phleboliths with a hyperintense T2-weighted signal, and marked enhancement suggestive of a pleomorphic adenoma but with poorly delineated contours. However, a hyperintense T1 signal can be misleading, suggesting cystadenolymphoma [11]. The presence of phleboliths, more difficult to visualize on MRI (hypointense signals) than on CT, is a good argument in favor of a diagnosis of venous malformation. In the present series, all of our patients had a hyperintense signal on T2 images with contrast injection. The T1 images were difficult to evaluate, giving a hypointense signal in six patients (60%) and a hyperintense signal in four (40%). Angiography is not helpful except in exceptional situations requiring preoperative embolization due to the size of the tumor or, in a postoperative context, if resection was incomplete.

US provides interesting information on arteriovenous malformations by visualizing a bundle of vessels with no tissue mass. The arterial bed is dilated with high Doppler velocity; arteriovenous microfistulae are also demonstrated. With angiography, a precise map of arteriovenous malformations can be obtained, demonstrating the main arterial blood supply. After this initial work-up and a multidisciplinary discussion, the decision may then be made to undertake endovascular embolization in preparation for surgical resection [12].

Cytology samples were obtained with needle aspiration in 30% of patients and were non-contributory in all cases. In the literature, needle aspiration remains the gold standard for the exploration of parotid tumors (in association with MRI), yet often fails to provide significant information. Moreover, it is clearly contraindicated in highly vascularized tumors due to the risk of bleeding [13].

In our present series, a definitive diagnosis of vascular malformation could not be achieved preoperatively in any of

the patients, even when the clinical presentation was highly suggestive. This can be explained by the fact that these tumors are very rare and that it is necessary to proceed by elimination to reach the diagnosis of vascular malformation. In addition, needle aspiration is not contributory and MRI is insufficient to formally exclude another histological type.

The question therefore becomes: What is the most appropriate surgical strategy? Do all vascular malformations require surgery, as these are theoretically slow-growing benign tumors with no risk of malignancy? In our series, all of the patients underwent surgery because the diagnosis could not be established preoperatively from just the imaging and needle aspiration findings. Histological proof was necessary even for the asymptomatic patients (50% of our series). The other 50% underwent surgery because of the pain caused by microthrombotic events and the increased volume (dyspnea or dysphagia) or aggressive nature of the lesion.

Treatment is mainly surgical because it allows a definitive diagnosis via histological examination. There is no proof of efficacy for complementary treatments such as radiation therapy. Ethanol injections have, however, been found to be a good alternative to surgery for venous malformations [14]. For very large lesions (requiring aggressive surgery), ethanol injection is an attractive alternative that can be associated with surgery in certain cases [15]. In our series, 90% of patients had undergone at least exofacial parotidectomy (only one patient underwent enucleation). For the two masseter tumors, the surgical approach required exofacial parotidectomy to allow exposure of the facial nerve branches for better dissection. Postoperative complications (paresis, Frey syndrome), but with complete recovery after a few months, were noted in 20% of patients. Only one patient had a recurrence despite complete resection at the first operation. The initial clinical presentation (fistula to the skin, vessel dystrophy, mixed vascular malformation) favored an aggressive tumor. On anatomopathological examination, the diagnosis was an arteriovenous malformation with poorly defined margins infiltrating neighboring tissues, but no evidence of malignancy.

Dissection and resection of vascular malformations is not a particularly complex procedure compared with other parotid tumors. Nevertheless, these tumors may occasionally be very advanced or particularly aggressive, thus requiring dissection of the facial nerve or even its total resection (to avoid recurrence). In this case, the surgical technique can be particularly complex, requiring surgical experience and skill.

In the literature, there have been only two reported cases of recurrence subsequent to incomplete resection due to poorly controlled bleeding [9,16].

As for histology, the classical appearance is benign vascular proliferation with endothelial cells in the vessel walls. The vessel lumen is either congested, or filled with thromboses or calcifications (phleboliths). Venous malformations have a regular encapsulated appearance and are bordered by regular-sized endothelial cells. Arteriovenous malformations have poorly defined margins with thin walls and variable-sized vessels. Direct communications between vessels that are arterial and venous in architecture are also observed [17]. In the present patients, the appearance was regular with a capsule in 90% of cases (all were venous vascular malformations). Only one patient had a non-encapsulated vascular malformation with irregular contours, the only case of a mixed arteriovenous malformation.

Conclusion

Vascular malformations of the parotid gland are rare entities; most involve venous malformations. The clinical course of these tumors distinguishes them from other vascular tumors such as hemangiomas, observed in children. The terminology currently uses the ISSVA classification based on clinical, evolutive, histological and hemodynamic data. The clinical presentation, needle aspiration and MRI are indispensable for making the diagnosis, which is generally, as in our present patients, difficult to establish. At present, imaging data, and especially MRI findings, offer the best preoperative diagnostic information. The gold standard treatment is surgical removal, especially as histological proof is required. Other therapeutic options are surgery-related or no treatment, with surveillance as an option if supported by clinical and imaging findings.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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