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Intraosseous venous malformation of the zygoma: A case report and literature review



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ABSTRACT

INTRODUCTION: Venous malformations of the zygoma are rare. Historically, venous malformations have been misrepresented as “hemangiomas”. The International Society for the Study of Vascular Anomaly (ISSVA) classification is a reasonable classification that leads to appropriate clinical diagnosis and treatment strategies. Collaboration between surgeons, radiologists, and pathologists is necessary for accurate diagnosis and management.

PRESENTATION OF CASE: We present here a case of an IOVM in a 59-year-old woman who was treated with a multidisciplinary approach. Superselective arteriography and embolization were effective for diagnosis as well as for prevention of large hemorrhage during surgery. En-bloc resection of the zygoma was performed within hours after embolization and autologous calvarial bone graft was used for primary reconstruction.

DISCUSSION: We performed a literature review consisting of reviewing 52 cases of IOVM of the zygoma discussing optimal material for reconstruction of the defect for intraosseous venous malformation of the zygoma nationally and internationally.

CONCLUSION: The combination of surgery and preoperative angiography makes it possible to prevent high risk of hemorrhage. For primary reconstruction of the zygoma, use of autologous calvarial bone can maintain the volume and reconstruct the natural malar contour.

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1. Introduction

Venous malformations of the bone of the maxillofacial region are rare. They account for less than 1% of all bony “tumors” [1,2,3] and are most frequently described in the vertebral column and calvarium [2]. The maxilla and mandible are the most commonly affected bones of the facial skeleton [3], followed by the zygoma, the orbit, and the condyle [2]. Fifty two cases of venous malformation (VM) of the zygoma have been reported in the literature [12,3]. Intraosseous venous malformation (IOVM) presents a rare yet unique clinical challenge to the surgeon. The keys for treatment are: (1) hemorrhage control, (2) en-bloc resection of the bony lesion including the normal bone, and (3) reconstruction after resection. Because of significant hemorrhage risk, intraosseous vascular anomalies can be life-threatening entities. Twenty five deaths by spontaneous hemorrhage have been reported in the literature [1]. In these literature, even the small lesions of VM may have significant hemorrhage risk. We present here a case of IOVM of the zygoma. Preoperative angiography was performed to confirm the diagnosis followed by embolization to reduce the risk of bleeding during the operation. A

good cosmetic result was achieved using autologous calvarial bone for reconstruction of the zygoma defect.

2. Presentation of a case

A 59-year-old female presented with a 3-month history of progressive painless swelling of the left cheek. A hard, immobile mass of the malar eminence was palpable (Fig. 1A). Computed tomography (CT) showed a round, well-defined expansile bony lesion measuring 5 cm in the left zygoma. The honeycomb lesion was also evident on 3-dimensional CT (3DCT) (Fig. 1B). Posteriorly, the lesion tapered into the anterior zygomatic arch. There were neither periosteal lesions nor any associated soft tissue lesions. The mass had an overall intermediate T1 signal intensity and a high T2 signal intensity on magnetic resonance imaging (MRI). Within the mass were areas of no signal that corresponded to the trabeculae seen on the CT study. No extra-osseous soft tissue component was apparent. On biopsy, significant hemorrhage was caused. The pathology revealed the lesion to be a cavernous hemangioma.

Preoperative superselective angiography and embolization were performed. Selective bilateral external carotid artery angiograms displayed markedly hypertrophied branches of the left facial artery and left internal maxillary artery (Fig. 2A). After microcatheter embolization, en-bloc resection of the zygoma including

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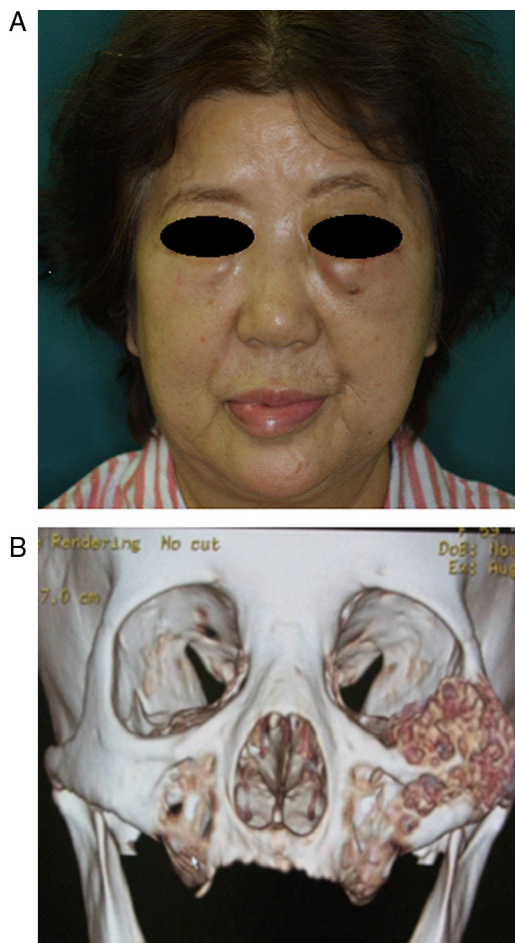


Fig. 1. (A) Preoperative view. (B) 3DCT scan image shows a honeycomb lesion in the left zygoma.

The lesion is a mass of mixed density over the malar eminence of the zygomatic bone involving the lateral orbital wall on the left side.

the VM was performed with a subciliary and intraoral approach to avoid postoperative facial nerve palsy caused by preauricular approach. The total blood loss was approximately 500 ml. Anatomical reconstruction of the zygoma and orbital floor was performed by using a 5 × 5 cm split parietal calvarial bone for the zygoma and a split iliac bone for the orbital floor. The split calvarial bone was taken primarily using a surgical chisel. The resulting calvarial bone donor site was repaired using hydroxyapatite bone cement.

Histology of the affected area (hematoxylin and eosin staining) revealed thin-walled, enlarged vascular channels with a single layer of flat, quiescent endothelium between bony trabeculae. The endothelium showed no signs of proliferation, mitotic figures, atypia, or tufting, and the bony margins did not show any sign of vascular anomaly.

Follow-up at 6 months demonstrated no significant deformity of the left zygoma, with good contour of the midface on clinical examination (Fig. 2B). CT scan at 6 month postoperatively, revealed volume maintenance and anatomical continuity of the calvarial bone grafts (Fig. 3). After over 3 years, the patient appears no recurrence or deformity of the left zygoma.

3. Discussion

A literature review of IOVM of the zygoma is shown in Table 1. Hemangiomas (Hs) and cavernous hemangiomas (CHs) are included types of IOVM. The incidence of intraosseous venous malformations of the zygoma occurs in a female to male ratio of 4.5:1. In 7% of cases, preoperative arterial embolization followed by total

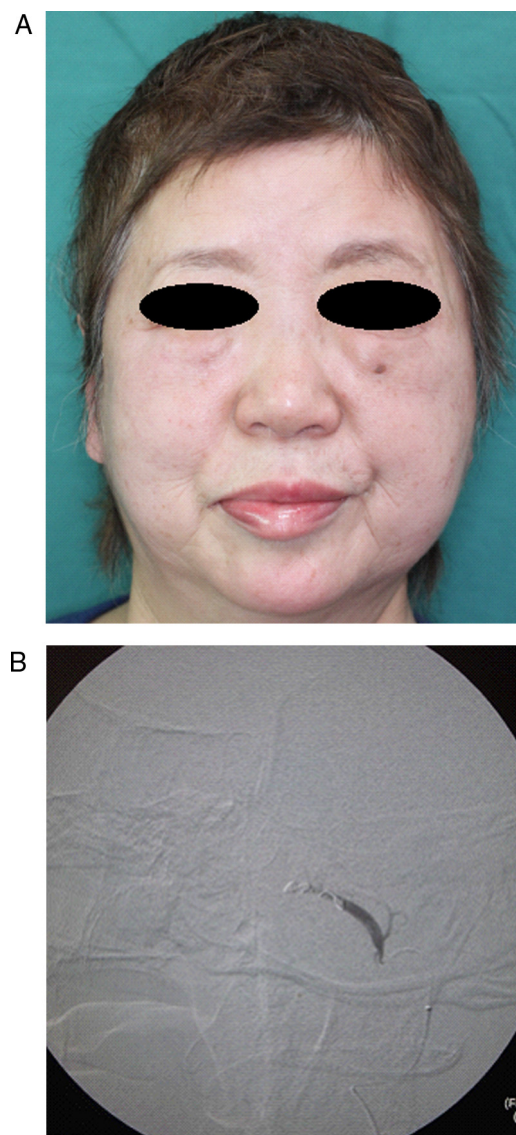


Fig. 2. (A) Lateral view of the selective left internal maxillary arteriogram before embolization. Homogenous staining was seen in the circle. The left internal maxillary artery was feeding the lesion. (B) Patient at 6 months postoperative.

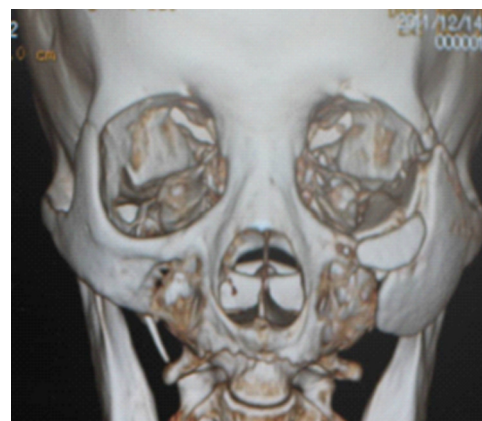


Fig. 3. 3DCT scan imaging 6 months postoperatively.

Table 1
Literature review of intraosseous venous malformations of the zygoma.

Authors	Year	Age/sex	Diagnosis	Treatment	Reconstruction
Schoefield[3]	1950	18 Month/male	H	Excision	NP
Sherman and Wilner[3]	1950	46 Year/female	H	Excision	NP
Walker and McHenry[3]	1950	40 Year/female	Capillary H	Excision	NR
	1950	10 Year/female	H	Excision	NR
Davis and Morgan[3]	1950	47 Year/female	CH	Ligation of external carotid A. and excision	Rib bone graft
Marshack[3]	1950	53 Year/female	Capillary H	Excision	Pedicled fatty tissue
	1950	35 Year/female	Capillary H	Excision and curettage	Layer of surgical
Schmidt[3]	1950	43 Year/female	Capillary H	Excision and curettage	Pedicled fatty tissue
Har-EL and Hadar[3]	1950	60 Year/male	CH	Excision	Silicone
	1950	43 Year/female	Mixed CH	Excision	Silicone layer
	1950	58 Year/male	CH	Excision	Silicone layer
Warman and Myssiorek[3]	1950	38 Year/female	CH	TR	Iliac bone
Jeter and Hockney[3]	1950	1 Month/female	CH	Ligation of external carotid A. and curettage	Surgical removed postoperatively
		50 Year/female	CH	Excision	Iliac bone graft
Clouser and Meneghini[3]	1991	56 Year/female	CH	TR	Split calv. bone
		35 Year/female	CH	TR	Split calv. bone
Cuesta Gil and Navarro-Vila[3]	1992	12 Year/female	CH	AE + TR	Split calv. bone
Savastano, Russo, and Aquila[3]	1997	41 Year/female	H	TR	Calv. flap
Hirano[4]	1997	83 Year/female	CH	TR	Hydroxyapatite
Yoshioka	2001	72 Year/female	CH	TR	Titanium mesh
Konior and Kelly[3]	1999	45 Year/female	MH	TR	Calv. bone
Moore and Chun[3]	2001	31 Year/female	H	AE + TR	Titan plate
Colombo et al.[3]	2001	75 Year/male	CH	TR	NS
Nakatani[16]	2003	9 Year/female	CH	TR	Rib bone graft
Koybasi, Saydam and Kutlay[5]	2003	33 Year/female	CH	TR	Hydroxyapatite
Fernandez, Luberti and Dominguez[3]	2003	26 Year/female	NS	TR or SR	NS
Ramchdani and Sabesan[3]	2004	38 Year/female	CH	TR	Pedicled calv. flap
Perugini, Renzi, and Beceli[3]	2004	60 Year/male	MH	TR	Calv. bone
		43 Year/male	Cellular H	TR	Calv. bone
		32 Year/male	MH	TR	Calv. bone
		46 Year/female	Cellular H	TR	Medpor
		32 Year/female	MH	TR	Medpor
		38 Year/female	Cellular H	TR	NR
Cheng et al.[19]	2006	50 Year/female	CH	PR/later TR	NP
Zins, Turegun, and Hosn[9]	2006	36 Year/female	CH	TR	Calv. bone P.O. 6 m
Riveros et al.[3]	2006	72 Year/NS	NS	TR	NP
Sakamoto et al.[8]	2007	47 Year/female	CH	TR	Ceratite
Gomez et al.[3]	2008	35 Year/female	IOH	TR	Calv. bone
Valentino et al.[6]	2008	57 Year/male	IOH	TR	Rib bone graft
		NS/NS	IOH	TR	Medpor/removal
		NS/NS	IOH	TR	Medpor/removal
		NS/NS	IOH	TR	Medpor
		NS/NS	IOH	TR	Calv. bone
		NS/NS	IOH	TR	Calv. bone
Srinivasan et al.[12]	2009	66 Year/female	VM	TR	NP
Zinz ¹⁷	2010	NS/NS	IOH	TR	Calv. bone
Defazio et al.[3]	2011	52 Year/female	VM	Surveillance	NP
		50 Year/female	VM	TR	Calv. bone
		49 Year/male	VM	TR	Harvested bone
Dhupar and Yadav[15]	2012	34 Year/female	CH	TR	NP
Leiboritch et al.[20]	2012	47 Year/female	IOCH	TR	NP
This case	2013	59 Year/female	IOVM	AE + TR	Calv. bone

H – hemangioma; CH – cavernous hemangioma; MH – mixed hemangioma; IOH – intraosseous hemangioma; TR – total resection; PR – partial resection; NP – not performed; and NS – not stated.

resection of the lesion was performed. With regard to reconstruction, 40% were with autologous calvarial bone graft and 40% were with non-autologous substances, such as silicone [3], polyethylene [3,6], titanium plate [3,7], or hydroxyapatite paste [4,5]. There was one recurrence after 6 months, and infection was reported in two cases that were reconstructed with hydroxyapatite [9].

3.1. Classification

Historically, the reporting of vascular anomalies has been misrepresented in the literature through the indiscriminate use of the term “hemangioma” to describe a variety of distinct vascular lesions [3,10,12]. This ambiguity has led to misconceptions and inaccuracies regarding the appropriate diagnostic approach and therapeutic management of vascular lesions [1,12].

The International Society for the Study of Vascular Anomaly (ISSVA) classification is a reasonable classification that leads to

appropriate clinical diagnosis and treatment strategies with the differentiation of vascular tumors and VM using simple, easily comprehensible nomenclature [10,12]. Although VMs have been described differently in the WHO classification as well as in the prominent textbook of Enzinger and Weiss, the ISSVA classification is the most logical and clear classification system [10]. Prior nomenclatures including cavernous hemangioma, venous hemangioma, intramuscular hemangioma, and synovial hemangioma are all categorized as VM in the ISSVA classification [10].

3.2. Etiology

VMs are malformations of the vascular system present at birth [1]. The frontal bone is the most common site (31%) followed by the temporal (13%) and zygomatic bones (12%) [11]. VMs are approximately 3–5 times more common in females than males [2,3,11].

3.3. Diagnosis

Zygomatic IOVMs may cause deformity of the cheek and orbit with proptosis and diplopia. Focal neurological findings and bruits are uncommon until late in their course [1].

The CT appearance most commonly shows a characteristic sharply marginated expansile lesion with intact inner and outer tables and a sunburst pattern of radiating trabeculae. “Soap bubble” and “honeycomb” configurations may also occur [2,12]. In the MRI, VM shows low to iso-intense areas within the muscle on T1 imaging with high intensity areas on T2 imaging [2]. In our case, the diagnosis of IOVM of the zygoma was suggested on the basis of the morphologic similarity of the lesion to a calvarial hemangioma. Intraoperatively, the lesions looked to be hard, blue–violet domed masses lying beneath the periosteum, and the bone seemed spongy.

Angiography is essential not only to confirm the diagnosis, but also to aid in the preoperative management of the tumors [13]. VMs often show dilations of the vascular supply and venous drainage, pooling of contrast in ectatic vascular spaces, and shunting. Because multiple arteries supply the lesion, embolization needs to be performed before the operation to prevent hemorrhage [13,14].

3.4. Treatment

Total en-bloc excision of bony lesions followed by reconstruction may be curative [3,4,12]. The indications for treatment include mass effect, high risk of hemorrhage, and cosmesis. Different types of treatments have been described including radiotherapy, embolization of the main afferent vessels, curettage, and resection [12,15]. Radiotherapy may stop the progress of the lesion but does not address the cosmetic deformity and has the risks of tissue necrosis, retardation of growth of bones and teeth, telangiectasias, and malignant degeneration [3,9]. Although embolization or ligation of the external carotid artery has been described, this strategy not only can lead to incomplete remission of the lesions, but also carries with it a risk of recurrence. En-bloc resection including a margin of normal bone around the lesion can decrease the recurrence and simultaneously avoid disastrous bleeding [1,6,7,9]. The honeycomb appearance of these lesions on CT is the result of reactive osteoblastic bone remodeling in response to the stress created by the enlarging VM, and the lesions are continuous with the normal surrounding bone. It is important to distinguish the margins of lesion in order to resect normal bone around it. The postoperative complications may be partial infection, disformity and recurrence. Temporary facial palsy has not found in among the literature regarding with IOVM of zygoma.

3.5. Reconstruction

The zygoma is critical to facial aesthetics. Reconstruction after resection is of great importance to preserve aesthetics and for ocular functionality of the zygoma orbital region. Primary reconstruction can maintain a normal bony foundation and prevent soft-tissue contraction [7,9]. Various reconstructive methods have been used including repair with a surgical pack [2,3], silicone implants [3], titanium mesh implants [3,7] hydroxyapatite cement [4,5], and autologous graft with rib and iliac bone [3,6,16]. We chose autologous parietal calvarial bone over other autologous bone grafts for primary reconstruction because the parietal curve has a similar contour to the zygomatic bone and also calvarial free bone grafts are well preserved in the recipient site, particularly in the face. Experimental studies have demonstrated enhanced volume maintenance for calvarial bone as compared with other traditional bone graft sources [9,17,18]. Because the ratio of cortical bone is higher in calvarial bone than in other bone grafts, such as

iliac and rib bones [17,18] and its dense cortical nature limits revascularization, the calvarial bone resorption rate is low [18]. Jinz [9] described that the use of autologous bone graft is better than using non-autologous materials like methylmethacrylate-based decalcified allograft bone or bone substitutes like hydroxyapatite cement because these can become infected or resorbed, particularly in the case of hydroxyapatite cement [9,17].

4. Conclusions

The field of vascular malformations has long been hampered by non-uniformity of language, which has frustrated attempts at research, diagnosis, and treatment. Recent advances in research reveal the underlying genesis of these lesions and have led to a coherent, reliable ISSVA classification system that facilitates uniformity in communication and efficacy of treatment. No single medical specialty in isolation can deliver high-quality care to these patients. Collaboration and mutual co-operation between surgeons, radiologists, and pathologists are necessary for accurate diagnosis and management. Superselective arteriography and embolization are extremely important diagnostic and therapeutic tool in arteriovenous and venous forms. The combination of surgery and embolization makes it possible to achieve definitive results. For primary reconstruction of the zygoma, the use of autologous calvarial bone can maintain the volume and reconstruct the natural malar contour via a relatively simple technique.

Conflict of interest

None.

Funding

None.

Ethical approval

The patient's approval has been given for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Toshie Matsumiya wrote the paper.

Hitoshi Nemoto analyzed the review.

Yoshiaki Kasai collected the literature for the review in the report.

Naoki Maruyama contrived the concept of this paper.

Guarantor

Hitoshi Nemoto would be the guarantor.

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