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REPORT, INTERESTING CASE

Intravascular Papillary Endothelial Hyperplasia of the Mandible in a 66 Year-Old Woman

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Abstract Intravascular papillary endothelial hyperplasia (IPEH) is a rare benign vascular non-neoplastic lesion that is characterized by papillary proliferation of vascular endothelial cells. Masson first described this lesion in 1923, and since then, it has been named Masson's tumor, intravascular angiomatosis, Masson's lesion, Masson's pseudoangiosarcoma, Masson's hemangioma, and vegetant intravascular hemangioendothelioma. The term IPEH is currently the most illustrative and the most frequently used in the literature. This lesion seems to occur in any blood vessel in the body, but it is more likely to be observed in the skin and subcutaneous tissues of the head and neck region, fingers, and trunk. Mandibular involvement is rare. IPEH's etiology remains unknown. Histopathological examination reveals a reactive proliferation of endothelial cells organized in minor papillary assemblies with hypocellular and hyalinized centers and arising from an organized thrombus. The appropriate treatment for IPEH is the excision with healthy margins; prognosis is good, with rare recurrences that may occur when the lesion is incompletely excised. The aim of the present article is to report and discuss the clinical, histopathological, immunohistochemical, and therapeutic issues of a case of IPEH of the mandible in a 66-year-old woman.

Keywords Intravascular papillary endothelial hyperplasia · Masson tumor · Diagnosis · Management · Mandible

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is a rare benign vascular non-neoplastic lesion that is characterized by papillary proliferation of vascular endothelial cells [1–10].

Masson first described this lesion in 1923, and, since then, it has been named Masson's tumor, intravascular angiomatosis, Masson's lesion, Masson's pseudoangiosarcoma, Masson's hemangioma, and vegetant intravascular hemangioendothelioma [3–12]. The term IPEH is currently the most illustrative and the most frequently used in the literature [3–12].

This lesion seems to occur in any blood vessel in the body, but it is more likely to be observed in the skin and subcutaneous tissues of the head and neck region, fingers, and trunk [1–8]. Mandible involvement is rare, with the first case being reported by Komori et al. [2] in the left mandibular body in 1984.

IPEH's etiology remains unknown. Nevertheless, it has recently been suggested as a reactive response to inflammation and stasis [1–12]. Histopathological examination reveals a reactive proliferation of endothelial cells organized in minor papillary assemblies with hypocellular and hyalinized centers and arising from an organized thrombus. The appropriate treatment for IPEH is the excision with healthy

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margins; prognosis is good, with rare recurrences that may occur when the lesion is incompletely excised [1–12].

To the best of our knowledge, only five cases of mandibular IPEH have been reported in the literature to date.

The aim of the present article is to report and discuss the clinical, histopathological, immunohistochemical, and therapeutic issues of a rare case of IPEH of the mandible in a 66-year-old woman.

Case Report

A 66-year-old woman had attended the Dentistry Department for years for the management of periodontal disease. Previous panoramic radiograph obtained in 2016 is depicted in Fig. 1. Past medical history included osteoporosis that was currently in treatment by monthly intramuscular injections of Clodronate 200 mg.

In 2022, the patient performed a panoramic radiograph to check the dental status (Fig. 2); it incidentally revealed a multilocular, expansile, well-circumscribed, osteolytic lesion occupying the right mandibular ramus.

Extraoral and intraoral physical examination did not reveal any abnormal finding or pathological sign; no intraoral swelling could be observed; no paresthesia or anesthesia were reported by the patient.

The patient underwent computed tomography (CT), which confirmed the presence of a multilocular, expansile, well-circumscribed, osteolytic lesion located in the right ramus of mandible, with lingual cortical erosion (Fig. 3).

A written informed consent was obtained from the patient and the patient underwent an incisional biopsy, and the material was sent to the pathologist for histopathological analysis. Histopathological findings supported the diagnosis of IPEH.

Then, the surgical excision under general anesthesia was decided, together with the patient; the mandibular lesion was completely removed via a vestibular approach.

Histopathological examination revealed intravascular proliferations of papillary processes lined with a single layer of endothelial cells, and fibrous materials. There were no significant nuclear atypia, hyperchromasia or mitotic activity. The endothelial cells were positive for

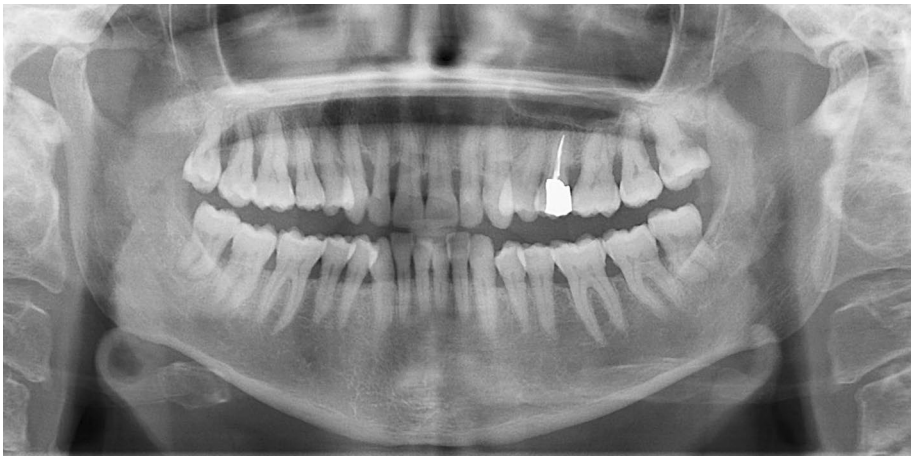

Fig. 1 Panoramic radiograph performed in 2016

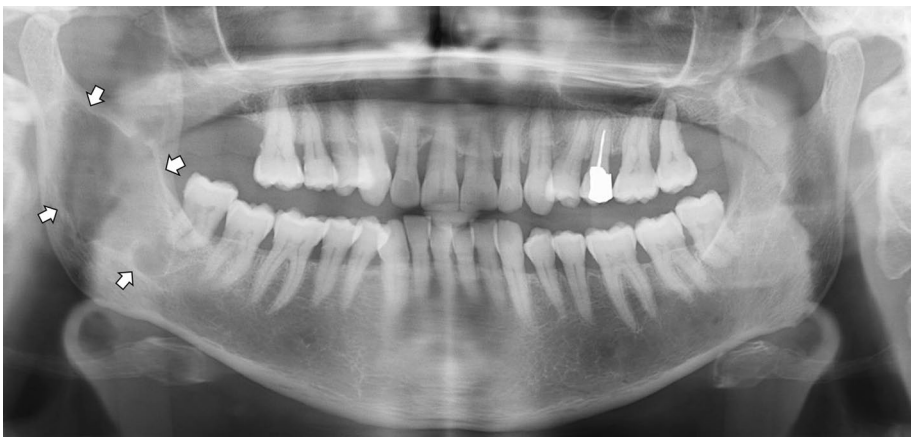

Fig. 2 Panoramic radiograph performed in 2022, revealing a multilocular, expansile, well-circumscribed, osteolytic lesion occupying the right mandibular ramus (the arrows points the borders of the lesion)

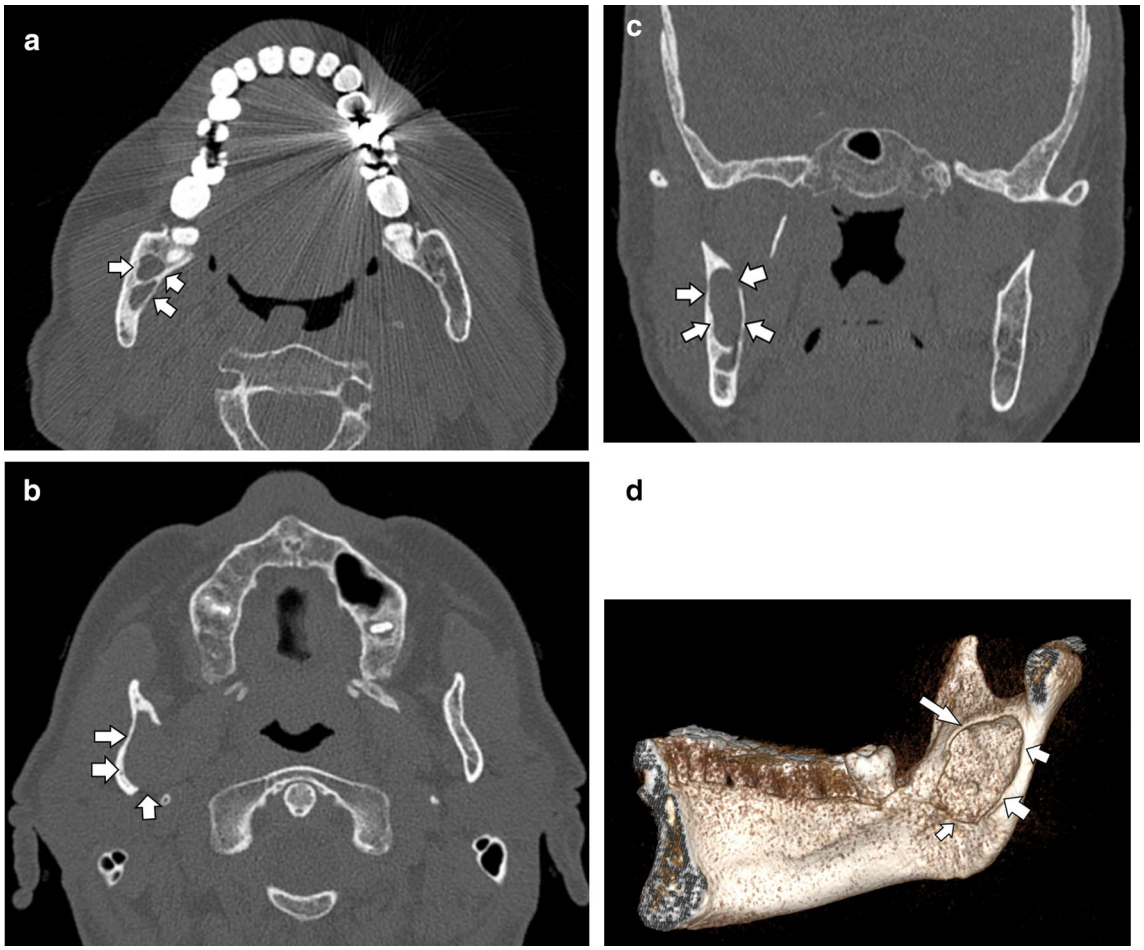

Fig. 3 CT scans (**A**, **B**, axial; **C**, coronal; **D** 3D) performed in 2022, confirming the presence of a multilocular, expansile, well-circumscribed, osteolytic lesion located in the right ramus of mandible, with lingual cortical erosion (the arrows points the borders of the lesion)

vascular markers CD31 and CD34. Histopathological diagnosis of IPEH was confirmed.

Postoperative course was uneventful.

Two years after surgery, no recurrences of the lesion were observed. Follow-up CT scan (Fig. 4) was obtained 9 months after surgery and showed the progressive healing of the right mandibular ramus.

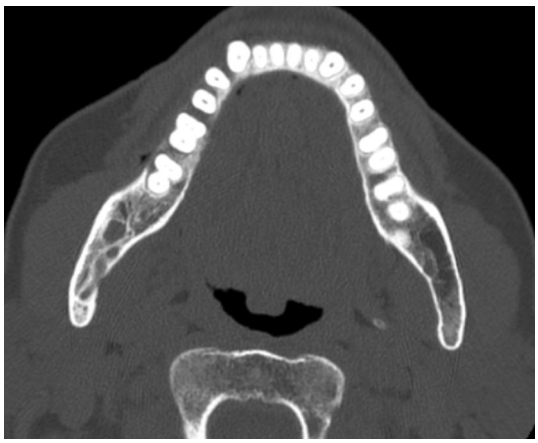

Fig. 4 CT scan performed in 2023, confirming the progressive healing of the right mandibular ramus

Discussion

IPEH is a benign, vascular lesion that appears to be rare in the oral region, with the lower lip being the most frequent site, followed by tongue, upper lip, and buccal mucosa. Mandibular site of IPEH is extremely uncommon, with just five cases previously reported in the literature [1–5, 8–11].

IPEH has been observed at any age from 9 months to 78 years with a slight female predominance.

Pathogenesis of IPEH is still unclear. An important role has been assigned to endothelial cell proliferation and

papillary formation in the vascular lumen that undergo degeneration and necrosis. Alternative proposed mechanisms have been a reactive process of endothelial cells induced by blood stasis and perivascular inflammation and a pseudotumoral lesion caused by endothelial proliferation with papillary formation preceded by an accumulation of thrombotic material [1–10].

Clinically, IPEH, when localized in soft tissues, usually manifests as a firm reddish-blue nodule or mass. Typical signs and symptoms are nonspecific and depend primarily on the anatomic location of the lesion. Furthermore, differential diagnosis may be challenging, as it is difficult to differentiate it from neoplastic and vascular lesions if only clinical findings are considered.

The differential diagnosis of soft tissue IPEH includes reactive, vascular and neoplastic lesions (hemangioma, lymphangioma, hematoma, Kaposi's sarcoma, hemangioendothelioma, thrombosed vein, traumatic fibroma, pyogenic granuloma, angiosarcoma, and salivary gland tumor). In patients with intrabony IPEH, differential diagnosis could include bony cysts and neoplasms, according to site [3–7]. In our case, the pharmacological anamnesis of the patient that was currently in treatment by monthly intramuscular injections of Clodronate 200 mg was also taken into consideration as for the vascular component proliferative action: nevertheless, the pathological examination allowed us to clearly establish a diagnosis of IPEH.

Radiologically, mandibular IPEH may present as an osteolytic, radiolucent, expansile, multilocular lesion with possible erosion of cortical plates. Bony septum and residual crests in the destructive areas may be observed, due to incomplete osseous destruction. Therefore, mandibular IPEH should be included in the radiological differential diagnosis of bone fibrous dysplasia, odontogenic keratocyst, ameloblastoma, intraosseous vascular tumors and malformations, and bone giant cell tumors [4–11].

In this case, panoramic radiograph incidentally revealed a multilocular, expansile, well-circumscribed, osteolytic lesion occupying the right mandibular ramus.

Computed tomography examination is crucial to highlight and assess the characteristics, the borders, and the expansile behavior of IPEH.

Anyway, it is difficult to diagnose IPEH by means of preoperative imaging examinations, thus making histopathological examinations necessary for a definitive diagnosis. Histologically, the pathognomonic feature of IPEH is the presence of papillary endothelial proliferations confined to intravascular spaces. Further typical histological features of IPEH include the association of most of papillary structures with thrombi; the fibrohyalinized tissue, lined with no more than two layers of endothelial cells, making up the papillae; the endothelial cells possibly being hyperchromatic, but with the absence of extreme nuclear atypia and frequent mitotic figures; and the absence of necrotic tissue [5–12].

Immunohistochemical staining for CD31 and CD34 has been reported to be positive in the majority of endothelial cells of IPEH lesions. In this case, endothelial cells were positive for CD31 and CD34: therefore, these findings were contributory to the definite diagnosis. Other markers can be used, such as factor VIII antigen, vimentin, laminin, ferritin, and alpha-smooth muscle actin. Nevertheless, different maturation degrees of the lesions may account for immunohistochemical staining variations. The histological differential diagnosis of IPEH includes angiosarcoma, spindle cell hemangioma, mucocele, hemangioendothelioma, intravenous pyogenic granuloma, Kaposi's sarcoma, malignant endovascular papillary angioendothelioma or Dabska's tumor, and intravascular endothelioma [2–11].

Three different types of IPEH have been reported: a primary (pure) form which is observable in a dilated vessel; a mixed form that occur in preexisting varices, hemangiomas, pyogenic granulomas, and lymphangiomas; and an uncommon type in an extravascular location [1, 3, 5, 9].

The treatment for IPEH is complete surgical excision. Some authors have proposed that preoperative arteriography could be performed together with a presurgical embolization in selected and large lesions to minimize the surgical bleeding. Prognosis of IPEH is excellent, and recurrence is extremely rare [1–8]. Our patient exhibited no evidence of recurrence during postoperative follow-up because of the complete surgical excision.

Commonly, intraosseous lesions may be overlooked because of their silent clinical development. In this case, an IPEH lesion was incidentally diagnosed following the execution of a routine panoramic radiograph. Furthermore, the previous negative panoramic radiograph performed 9 years (Fig. 1) and 6 years (Fig. 2) before, respectively, shows the potential for a rapid progression by IPEH.

Conclusions

It is fundamental that dental practitioners are aware of clinical, radiological, and histological features of IPEH in order to include this pathological condition in the differential diagnosis of intrabony and soft tissue lesions.

Author Contributions MM contributed to conceptualization; MM and PB provided methodology; AG and MM performed validation; PB and MF carried out formal analysis; PB and MB performed data curation; PB performed writing—original draft preparation; PB, MF, and MM performed writing—review and editing; MB and MM performed supervision. All authors have read and agreed to the published version of the manuscript.

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