

CASE REPORT

Axis fracture due to giant cranial AVM

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Abstract

Cerebral arteriovenous malformations (AVMs) are a vascular anomaly consisting of a bundle of direct connection of arteries and veins. AVMs clinical expression ranges from complete asymptomatic, and thus incidentally found, to life threatening with rupturing and bleeding. In this wide spectrum, osteolysis is considered as a rare complication of interosseous AVMs, and only few cases of mandible and maxilla osteolysis have been reported. We present, herein, a case of an intracranial AVM, which has caused in the course of the time an osteolysis of the dens and axis.

INTRODUCTION

Brain arterial malformation (AVM) belongs to cerebral vascular diseases and constitutes an abnormal direct connection between arterial and venous system in absence of an intervening capillary network. Due to this defected structure, arteriovenous malformations (AVMs) are prone to rupture, causing intracranial hemorrhage. Its clinical course can widely vary from asymptomatic, and thus only incidentally diagnosed, to life-threatening conditions. Within this wide spectrum, an AVM may present with headache, seizures and long-term disability [1].

The gold standard to diagnose an AVM is through angiography, but thanks to the recent development in noninvasive imaging techniques, the incidence of asymptomatic AVMs has consecutively increased [1].

Osteolysis as a complication of an AVM has only rarely been reported and has predominantly been associated with interosseous AVMs. Additional manifestations of an interosseous AVM may be pain, pulsating mass, regional bone overgrowth and skin ulceration [2]. Interosseous AVMs themselves constitute a very rare condition mostly present in the craniofacial



Figure 1: Noncontrast CT of the cervical spine reveals the unstable odontoid fracture at the basis of dens. This fracture is classified as Type II on Anderson and d'Alonzo classification. Despite the fall incident, this fracture should be taken into account as a pathologic fracture. The osteolysis and the sclerosis of the fracture margin is caused by a mass inside the bone, revealed to be an AV malformation reaching from the right side of the axis to the basis of dens.

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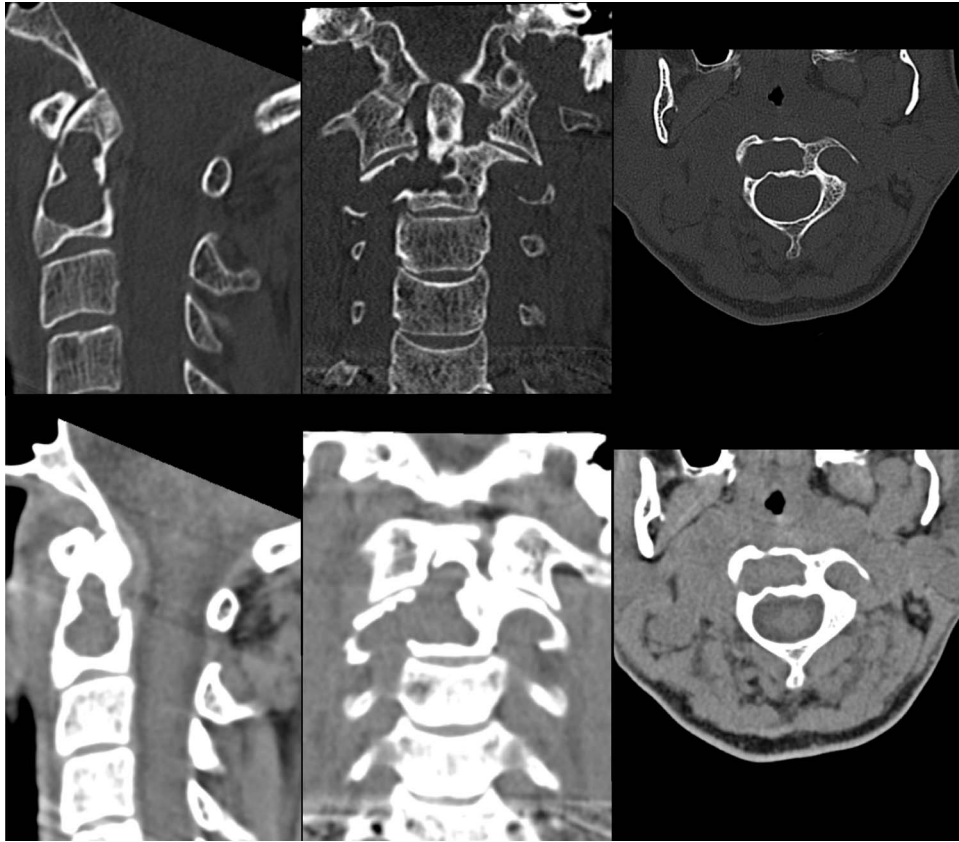


Figure 2: Noncontrast CT of the cervical spine shows the extended bone destruction likely on the ground of preexisting AV malformation.

region with primary involvement of the mandible and maxilla [3].

CASE REPORT

A 52-year-old woman presented in our neurosurgical emergency ward with severe headache accompanied by nonspecific radiating neck pain. The headache persisted for 3 days, and due to an exacerbation, the patient was admitted to the hospital.

Medical history revealed that the patient fell in the last few weeks due to a known right-sided hemiplegia. The physical examination did not show any sign of external injury like abrasions, hematomas, etc. The neurological physical examination showed an oriented but aphasic patient with a preexisting high-grade right-sided hemiparesis (3/5). The aforementioned hemiparesis resulted from an earlier (around 1976) hemorrhage of an AVM on the left hemisphere with accompanied intraventricular bleeding. The pupils were equal in size, reactive to light, and there was no relative afferent pupil defect. Moreover, an exophthalmus of the left eye was revealed as well as a known strabismus divergens and a facial nerve paresis on the right side. Furthermore, signs of meningeal irritation in form of a stiff and painful neck were detected. A brain computed tomography (CT) imaging was conducted to investigate any intracranial pathology responsible for the headache symptoms as well as the hemiparesis. The results of the CT imaging were similar to those of 10 years ago, detecting no pathological findings. Thus, we proceeded with a cervical spine CT scan due to the accompanied cervical symptoms. The latter revealed an unstable fracture

through the base of the dens, type II based on the Anderson and d'Alonzo classification [4]. There was no evidence of a spinal hematoma. Moreover, an extended osteolysis was revealed with reactive sclerosis of denses and axis' endplate and margins, as shown in Figs 1 and 2. The dens fracture was evaluated as a pathologic fracture. The fall incident could not explain the expanded osteolysis. Our assumption, based on the conducted imaging processes, is that the fracture was caused by a large AVM, which extended from the right side of the axis till the basis of the dens. The AVM also spread into the surrounding soft tissue.

During the hospitalization period, the neurological status of the patient remained unchanged, with persistent mobility difficulties. Taking into consideration the giant craniocervical AVM, the osteolysis of dens coupled with the decreased general condition of the patient, as well as her wish to avoid any kind of treatment, a conservative approach was chosen. The conservative treatment involved the adjustment of analgesics, a stiff neck collar for at least 6 weeks and regular application of physiotherapy.

DISCUSSION

Here, we publish a rare case of an osteolytic lesion caused by a giant AVM. Up to date, only a few case reports exist, describing a comparable condition. Our assumption is that the giant AVM gradually infiltrated the adjacent vertebral body causing osteolytic lesions over years. This assumption is based on the fact that the osteolysis was a previously unknown radiological

finding of a known AVM and that the fall incident could not explain this finding. Due to the patient's rejection of further diagnostic and therapeutic procedures, an angiographic or histological confirmation of our assumption was not possible. For the same reason it was impossible to clarify the precise localization of the AVM. The lack of a comprehensive diagnostic control in order to exclude other types of osteolytic lesions, such as tumors, sets a limiting condition. Angiography remains the gold standard of depicting an AVM and cannot be replaced by the advanced imaging technique of CT.

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CONFLICT OF INTEREST

None declared.

FUNDING

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ETHICAL APPROVAL

Ethical approval was not required based on the Journal Policies.

CONSENT

In respect of patient's privacy, identified information are not to be published. The patient and the relatives have been informed about the content above. The written consent was obtained.

GUARANTOR

K.L. is the guarantor for this manuscript.

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