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Dear Editor,
I have just sent you a revision manuscript.
We will change the form (from a case report to a Letter to Editor) and resend you the correct form of author declaration.

Best regards,
Dr. G. Pilloni

TITLE PAGE

Intramuscular myxoma of the cervical muscles with GNAS mutation

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TEXT

Intramuscolar myxoma (IM) represents a benign rare tumor, presenting more frequently in thighs, upper limbs and shoulder girdles.[1;2] In Mazabraud's syndrome (MS) multiple intramuscular myxomas are associated to monostotic or polyostotic fibrous dysplasia. MS represents a rare genetic disease characterized by the mutation of GNAS gene. [2] To our knowledge only four cases of myxomas localized in the cervical muscles are described in literature. No article described IM associated with spinal canal involvement.

We report the case of a 69-year-old man presenting with a painless posterior cervical mass increasing in size slowly during the past 5 years.

The mass was pulseless and strictly adherent to the deep tissues. The overlying skin was normotrophic. The patient complained ipoesthesia in the suboccipital region. A Magnetic Resonance (MRI) was performed showing an oval, solid intramuscular mass, measuring 5 cm. The lesion was hypointense in T1-weighted sequences and hyperintense in T2, with no intralesional calcification. (Figure 1) A tru-cut biopsy was performed with a pathology report of intramuscular myxoma with GNAS mutation. A CT scan with iodine contrast medium showed a solid, nodular and capsulated lesion measuring 50x44x40 mm. It was strictly adherent to C1 arch and the odontoid process, with splenious, inferior obliquus capiti, longissimus capitis, and semispinalis capitis muscles dislocation. It was localized next to the extraforaminal tract of the right vertebral artery (Figure 2). Neurologic examination on ward admission was normal, except for ipoesthesia in suboccipital region and a mild anisocoria (left pupil larger than the right one). The mass was surgically excised in general anesthesia with the patient in prone position. The head was locked in Mayfield head holder in moderate neck flexion. A longitudinal posterior incision gave access to C1 tubercle. Into the deep muscular plane a solid mass with a grey-whitish coloured capsule was evident. The lesion was strictly adherent to the vertebral artery. Considering the benign nature of the lesion and the proximity of the vertebral artery the capsule was opened. Multiple fragments of solid fibrous material (Figure 3) were excised and sent for pathology with the external capsule. The lesion was totally removed using an ultrasound knife (Figure 4) without any damage to the vertebral artery as confirmed by the intraoperative use of a doppler probe. (Figure 5) The pathology report confirmed the diagnosis of benign intramuscular myxoma. Patient was discharged after seven days. Considering the presence of GNAS mutation the patient was accurately informed of the possible diagnosis of MS and a 99-Tc Bone Scan was proposed to exclude the presence of a monostotic or poliostotic fibrous dysplasia. The patient, totally asymptomatic elsewhere, refused to perform the examination also considering the benignity of the diagnosis.

Follow-up at 6 months showed complete resolution of preoperative symptoms and no evidence of local recurrence at the MRI.

Myxoma is a benign mesenchymal tumor characterized by undifferentiated cells in a myxoid stroma. Lesions originated from skeletal muscles, are called IMs. Intramuscular myxoma represents a rare tumor with an incidence of one case per million of population per year. [3]. MS is an even more rare condition with only 81 cases reported at present. [2]

The most frequent sites for myxomas are represented by thigh, shoulder, buttock, and arm; lesions originated by lumbar back muscles are rare. IM is usually asymptomatic, pain is described only in 20% of cases

Nowadays, only sixteen cases are reported in the literature and only four of these cases raised from cervical paraspinal musculature [4]

At the MRI the IM appears like a cystic homogeneous mass, without contrast medium enhancement. On T1-weighted sequences the signal has high intensity, while on T2-weighted images it is brighter than fat. [5]

In some cases the lesion could infiltrate muscles. It is therefore important to make a differential diagnosis with other possible malignant neoplastic lesions (myxofibrosarcoma, fibromyxoid sarcoma). A preoperative biopsy is mandatory to decide the adequate surgical approach, whether wide or marginal/intralesional complete excision.

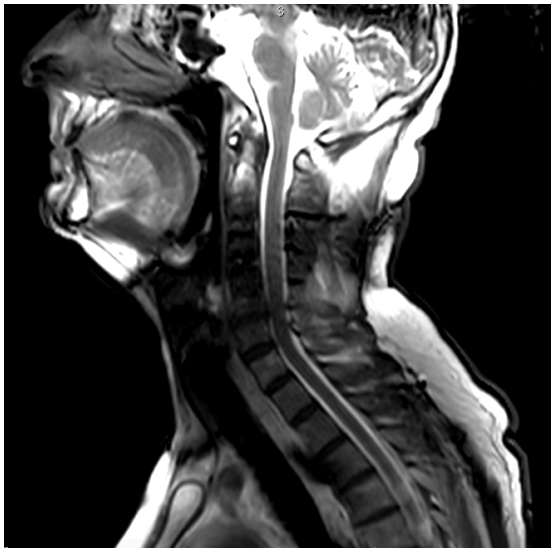
IM should be included in differential diagnosis of cervical paraspinal tumors. Total tumor excision is indicated due to the high risk of local recurrence reported after subtotal resection.

REFERENCES

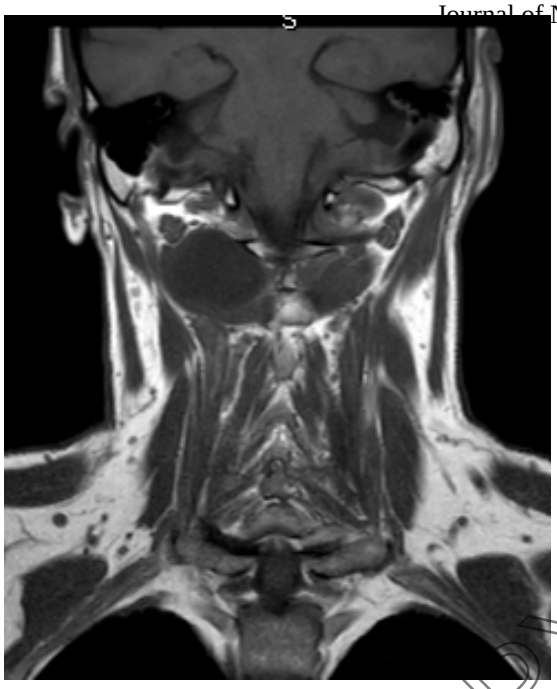
1. *Pérez Sánchez P, González Llorente J. Mazabraud's syndrome, an uncommon association of intramuscular myxoma with fibrous dysplasia. Radiologia. 56(3):281-3, 2014*
2. *Hashimoto H, Tsuneyoshi M, Daimaru Y et al. Intramuscular myxoma. A clinicopathologic, immunohistochemical, and electron microscopic study. Cancer. 1;58(3):740-7, 1986*
3. *Miettinen M, Hockerstedt K, Reitamo J et al. Intramuscular myxoma—a clinicopathological study of twenty-three cases. Am J Clin Pathol;84:265–72, 1985*
4. *Tataryn Z, Tracy J, Tsang C, Wu J, Heilman CB, Wein RO. Intramuscular myxoma of the cervical paraspinal musculature: case report and review of the literature. Am J Otolaryngol. Mar-Apr;36(2):273-6, 2015*
5. *Abdelwahab AF, Kenan S, Hermann G et al. Intramuscular myxoma: magnetic resonance features. Br J Radiol 65:485–490, 1992*

Figures captions

- *Figure 1.Pre-operative MRI: T2 sagittal image weighted, (A),coronal T1 image weighted (B),axial T1 weight-image with contrast enhancement (C)*
- *Figure 2. Pre operative Angio-CT scan to study vertebral artery course, strictly adherent to the tumor.*
- *Figure 3. Capsule incision*
- *Figure 4. Tumor total resection with ultrasound knife*
- *Figure 5. Ultrasound doppler probe to evaluated the vertebral right artery integrity after tumor resection*



Figures 1 A



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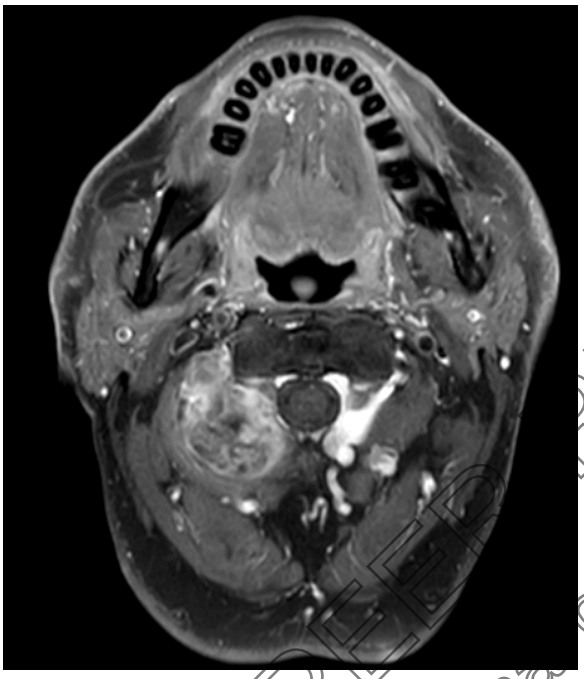

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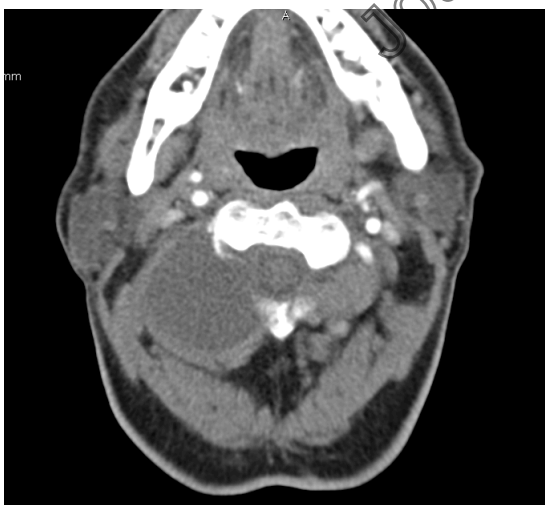

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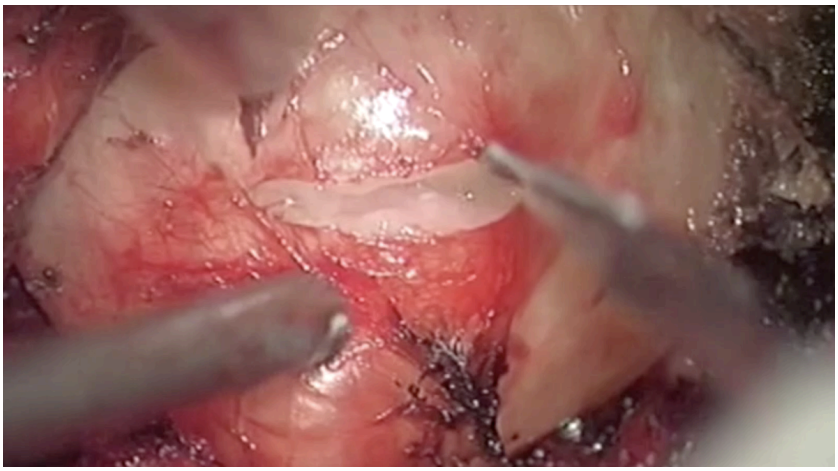

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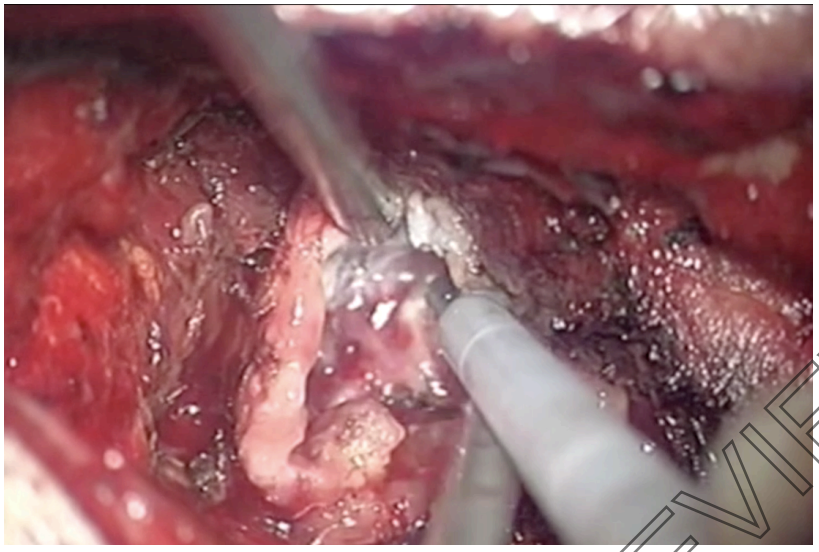

Figure 4

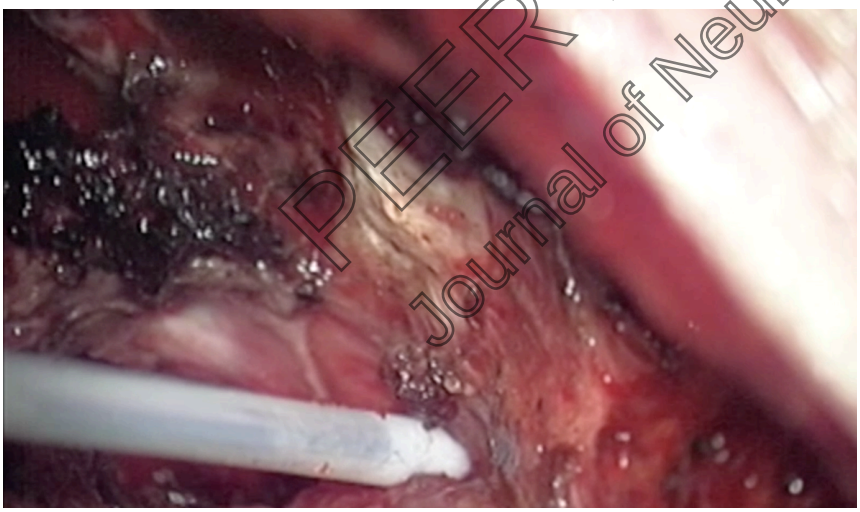

Figure 5