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Case report

Sacral solitary fibrous tumour: surgery and hadrontherapy, a combined treatment strategy☆

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1. Introduction

Hemangiopericytoma (HPC), recently associated by the WHO classification with Solitary Fibrous Tumor, is a kind of soft tissue sarcoma. It originates in Zimmerman's pericytes,[1] i.e. spindle cells wrapping the capillaries and postcapillary venules that provide mechanical and functional support to the endotheliocytes. Because of its origins, HPC can occur wherever endothelial tissue is present, the most common sites being the lower extremities, retroperitoneum and head-and-neck region.[2] It accounts for less than 1% of all vascular tumors and 5% of all sarcomatous tumors. Not only can this tumor have a locally aggressive behavior, especially inside the nervous system, it can also recur locally and metastasize through the bloodstream, mainly in the lungs and bone.[3] HPC can affect equally both sexes, mainly in adults in the fourth through sixth decades of life. It can manifest itself in two forms: soft and osseous with a slight predominance among males. The cellular architecture is characterized by tumor cells arranged in sheets and fascicles with staghorn-shaped vessels and dense intercellular reticulin meshwork (particularly visible at the silver staining) without evidence of epithelial membrane antigen (in contrast to meningiomas). At immunohistochemical study the tumor is positive for Vimentin, CD34 but negative to Factor VIII, S-100 protein, Actin, CKP, EMA and Desmin3. The presence of necrosis, hemorrhage, anaplasia and a high mitotic activity (more than five mitoses per ten microscopic fields), are a poor prognostic factor.[2] Surgical resection is the treatment of choice, with the use of chemotherapy reserved for metastatic or non-resectable lesions, albeit with no standardized regimens. Radiotheraphy, too, may play a role, as a sole management for lesions deemed not viable for surgery or on remnant/recurrencies.[4]

We describe a particular case of a sacral HPC, a rare form of this tumor (nine cases described in literature so far[2,3,5–11]), surgically treated in two steps: the first being a laparotomic approach whereas the second via a posterior approach by laminectomy. Since a small residual part was to be found after surgery, our treatment plan also included an innovative approach with Hadron Therapy (HT) in order to obtain tumor control while potentially limiting local side effects.

2. Case description

A 73-years-old female patient came to our observation as a result of investigations for a persistent left sciatica. A following MRI of the lumbar spine showed the presence of a sacral expansive lesion at the S1 level, with pre-sacral extrinsication ($60 \times 40 \times 80$ mm in dimension) (Fig. 1). In anamnesis we reported hypertension, total hysterectomy and bilateral saphenectomy. The patient suffered from hypoesthesia in the S1 area of the left inferior limb, without focal neurological deficits. The first surgical step was performed via an anterior laparatomic approach with the aid of a

☆ We report the case of a 73 year-old female with a sacral hemangiopericytoma treated surgically and with hadrontherapy. In this paper, we discuss the importance of a combined treatment strategy for this kind of lesion and the role of hadrontherapy.

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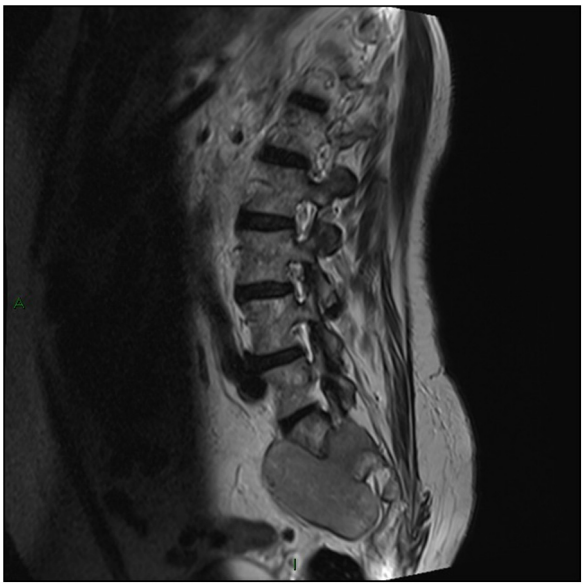

Fig. 1. Preoperatve sagittal T1 MR.

general surgeon. After partial mobilization of the sigma and the rectosigmoid junction for the left side of the pelvis to be exposed, the neoformation was seen strictly attached to the local structures. In particular, the ipsilateral hypogastric vein, from which the lesion was gently detached, had a minimal rip, promptly repaired. The tumoral tissue was then carefully separated from the sacral plate. Vascular plexi were to be found on the interface with the sacral bone, the lesion being highly vascularized. With the aid of both a Cavitron Ultrasonic Surgical Apirator (CUSA) and a microscope, we were able to resect most of the anterior part of the lesion. The post-operative course was regular and without complications. The histological examination reported micro neoplastic proliferation with solid architecture, elongated and angulated vascular channels, consisting of small-size elongated elements, with poor cytoplasm and round or oval nuclei, with dispersed chromatin; fine reticulinic texture in the absence of significant deposition of collagen. The neoplastic elements were immunoreactive for vimentin, BLC2 and CD34, negative for S100, cytokeratin, leiomuscolar markers and CD99. The Ki67+ proliferative index was about 5% and the mitotic index was 1 mitosis per 10 high-magnification fields. No necrosis was found. All these elements were compatible with the diagnosis of HPC (Fig. 2). A post-operative MRI control at 1 month showed the presence of a left sacral remnant at the S1–S2 level (Fig. 3). The second surgery was performed via a laminectomy of S1: a medial displacement of the dural sac by the lesion was observed in addition to a diastasis of the L5 and S1 nerve roots.The tumor was excised as much as possible until the relaxation of the dural sac and of the nerve roots. The post-operative course was without any complications. A post-operative MRI at 1 month showed a small residual tumor in the sacrum (Fig. 4). After a multidisciplinary team evaluation between Neurosurgeons and Radiation oncologists, the patient received a radiotherapy treatment with carbon ions on the residual sacral lesion at level S1–S2. The radiotheraphy took place at the Centro Nazionale di Adroterapia Oncologica (CNAO) in Pavia. The radiotherapy plan (Fig. 5) included a total dose of 73.5 Gy (RBE), (4.6 Gy (RBE)/irradiation, 16 irradiations/1 month, 4 irradiations/week), with SBO (single Beam Optimization)/IMPT (Intensity Modulated Particle Therapy) technique. The global treatment plan (2 surgeries + RT) ended 1 year after the onset of symptoms. On the first follow-up appointment (about 5 months after the radiotherapy), the patient had pain regression and no more paresthesias. Treatment was well tolerated and no interruption was needed, the patient experienced grade 1 erythema (according to Common Terminology Criteria for Adverse Events Version 4.0). 6 months later, a control MRI scan reported a 1-cm alteration of the posteroinferior edge of the L5 body, with fat density, probably representing a degenerative process. So far, i.e. after approximately 22 months from the radiotherapy treatment, she is clinically and radiologically stable (Fig. 6).

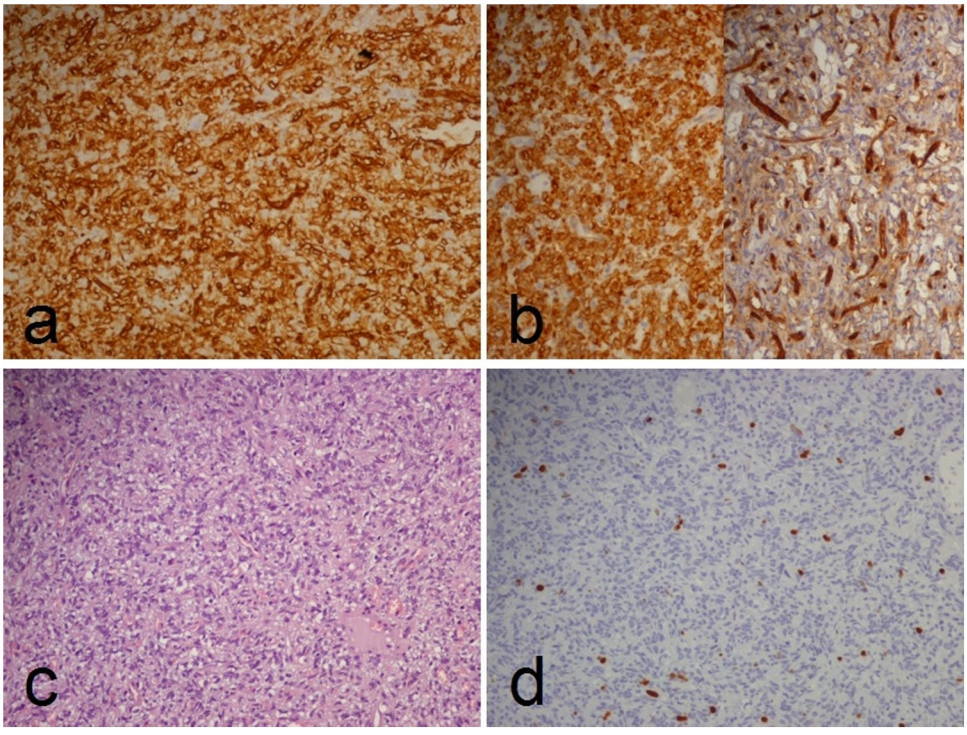

Fig. 2. Histologic images: vimentin (a), BLC2 and CD34 (b), HE 20× (c) e Ki67 (d).

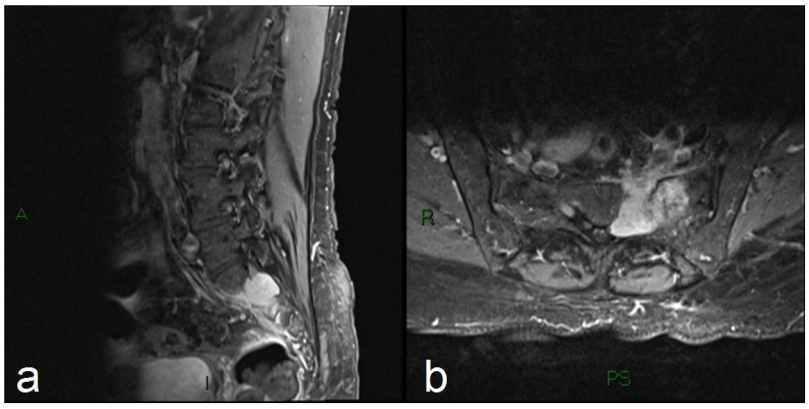

Fig. 3. Sagittal (a) and axial (b) T1 with gadolinium first control MR.

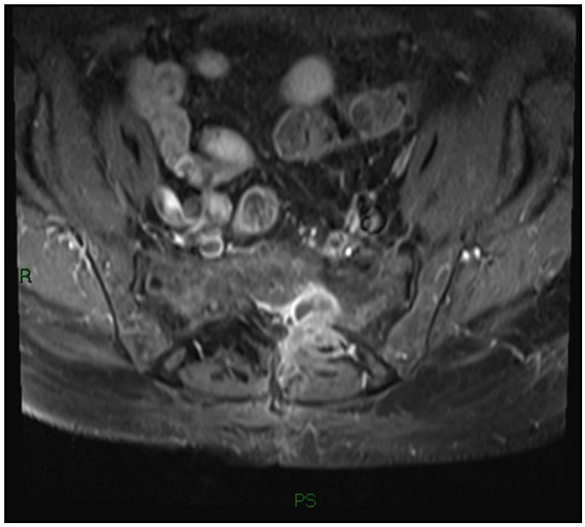

Fig. 4. Axial T1 with gadolinum second control MR.

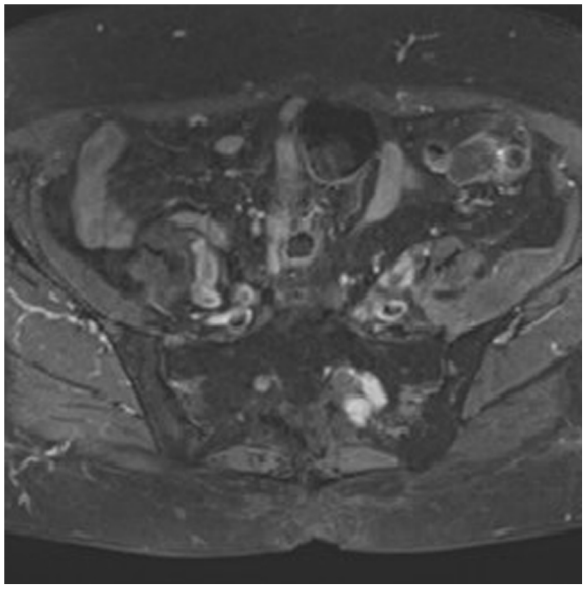

Fig. 5. Radioterapy plan.

3. Discussion

HPC was reported for the first time by Stout and Murray,[5] and albeit no clear etiology is known, some reports have pointed out a relationship with chemicals exposure, as herbicidal and vinyl chloride.[3] The most common symptoms at presentation are pain and palpable mass.[6] Particularly in sacral localizations, it may cause radiating pain as well as reach remarkable volumes before the onset of the symptoms.[2] That is why their treatment may be difficult and may need to be multidisciplinary. In particular the presence

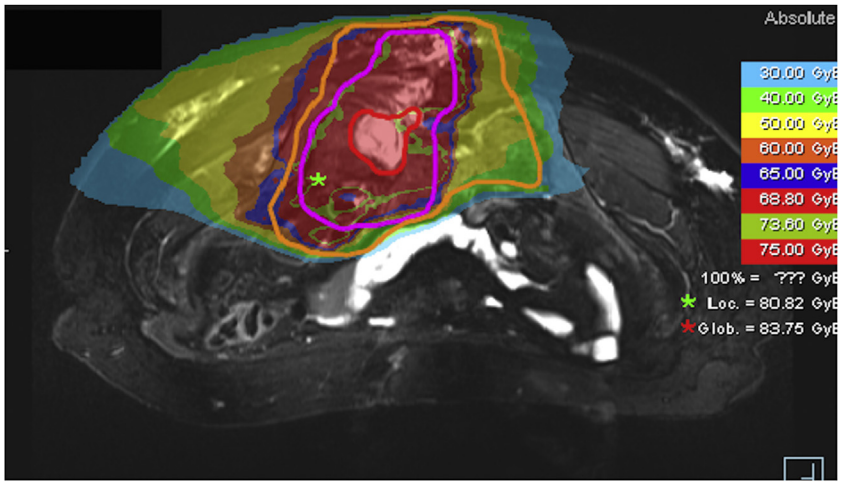

Fig. 6. Axial T1 with gadolinum last control MR.

of high-flow vessels into the lesion may be managed by means of pre-operative embolization.[12] Moreover, chemotherapy may sometimes play a role: the CYVADIC protocol (cyclophosphamide, vincristine, adriamycin and dacarbazine) is used as therapy for advanced, malignant lesions.[2] If applicable, neoadjuvant radiotherapy has been reported as useful to turn a non-resectable lesion into a resectable one, whereas adjuvant radiotherapy may improve the survival rate.[13]

HPC has usually 2- and 5-year overall survival rates of 93% and 86%, respectively. Nevertheless, in view of both its late local recurrences (4%) and distant metastases (20%), a follow-up is strongly recommended in the long run. All these considerations led our multidisciplinary team to treat the lesion as radically as possible while trying to preserve the physiological functions of the area, with particular regard to the nervous structures. Surgically-wise we do not damage the main nerve roots involved in the lesion and the presacral nerve structures known as hypogastric plexi. Given the proximity to the lesion treated with both these structures, our team envisioned that the use of HT could be a good compromise between tumor control and nerve-sparing. As a matter of fact, HT has a favorable dose profile, thus allowing the administration of high doses to the target while minimizing the dose to the surrounding tissues.[14] Moreover, the experience with sacral chordomas has already showed a good safety profile for this treatment in that region. One of its potential advantages over other RTs could therefore be its lower incidence of side effects/damage to surrounding structures. We must underline, though, that our report is a preliminary experience and must be considered as such. Further studies are needed to truly evaluate the effectiveness and potential advantages of HT in the combined treatment of sacral HPCs.

4. Conclusion

HPC of the sacrum is a very rare lesion. A combined surgical-radiotherapeutical treatment strategy may be needed. Given its safety profile, HT may have a role in this kind of tumors.

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Conflict of interest

None declared.

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