

A patient with sinonasal leiomyoma presenting with exophthalmos: Case report and review of the literature

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Abstract

A 71-year-old woman presented with a 2-month history of progressive exophthalmos. Magnetic resonance imaging revealed a tumor occupying the right frontal sinus that compressed the right orbit and eye ball. The tumor was resected through a right frontal craniotomy. The pathologic diagnosis was leiomyoma and similar to the histologic diagnosis of a uterine leiomyoma resected 4 years earlier. We suggest that this rare sinonasal leiomyoma was a benign metastasizing leiomyoma from the uterus to the nasal sinus.

INTRODUCTION

Leiomyomas are benign myogenic tumors that most commonly arise in the uterus. Other organs lacking smooth muscle are rarely involved and neck and head lesions are particularly rare. To our knowledge, only 10 sinonasal leiomyomas have been reported in English literature to date.^{2,7,8,16}

Benign metastasizing leiomyoma (BML) was first documented in 1939.⁵ The term BML has since been used to describe multiple benign leiomyomatous lesions in patients with a previous history of leiomyoma of the uterus. We present here a patient with BML to the nasal sinus.

CASE REPORT

The patient is a 71-year-old woman with 2-month history of progressive right exophthalmos. Her past medical history included resection of a uterine leiomyoma 4 years earlier. She had difficulty opening the affected eye, whose motility in all directions was restricted.

Computed tomography (CT) revealed a 30-mm-diameter tumor involving the right frontal sinus with erosion of the frontal bone (Figure 1a). Magnetic resonance imaging (MRI) showed an inhomogeneously enhanced tumor in right frontal sinus. It was isointense to the brain on T1- and mixed iso- and hyper-intense on T2-weighted images. The tumor compressed the right levator palpebrae superioris-, superior rectus-, and

superior oblique muscle, and the eye ball (Figure 1b-f).

The sinus was transcranially opened and an elastic grayish-red tumor was removed without resulting in major bleeding (Figure 2). There was no tumor invasion into the orbit or brain; the dura mater and periorbital membrane were intact. Postoperative MRI confirmed its total removal (Figure 3a-c).

Histologically the tumor composed of close spindle-shaped smooth muscle cells with elongated nuclei. The tumor cells manifested mild nuclear atypia. Immunohistochemically they were positive for α SMA, desmin, and estrogen receptor and progesterone receptor (Figure 4a-e). Based on these findings we made a diagnosis of sinonasal leiomyoma, a BML to the nasal sinus from the uterus. Postoperatively her exophthalmos improved dramatically and eye movement in all directions improved 3 weeks after surgery.

DISCUSSION

Leiomyomas are benign myogenic tumors that most commonly arise in the uterus (95%), skin (3%), and gastrointestinal tract (1.5%).^{3,5,10} They are rarely seen in other organs that lack smooth muscle, including the nasal sinuses. BML occurring years after hysterectomy is a rare entity seen in women aged 35-55 years.¹¹ Although there have been reports of morbidity and mortality

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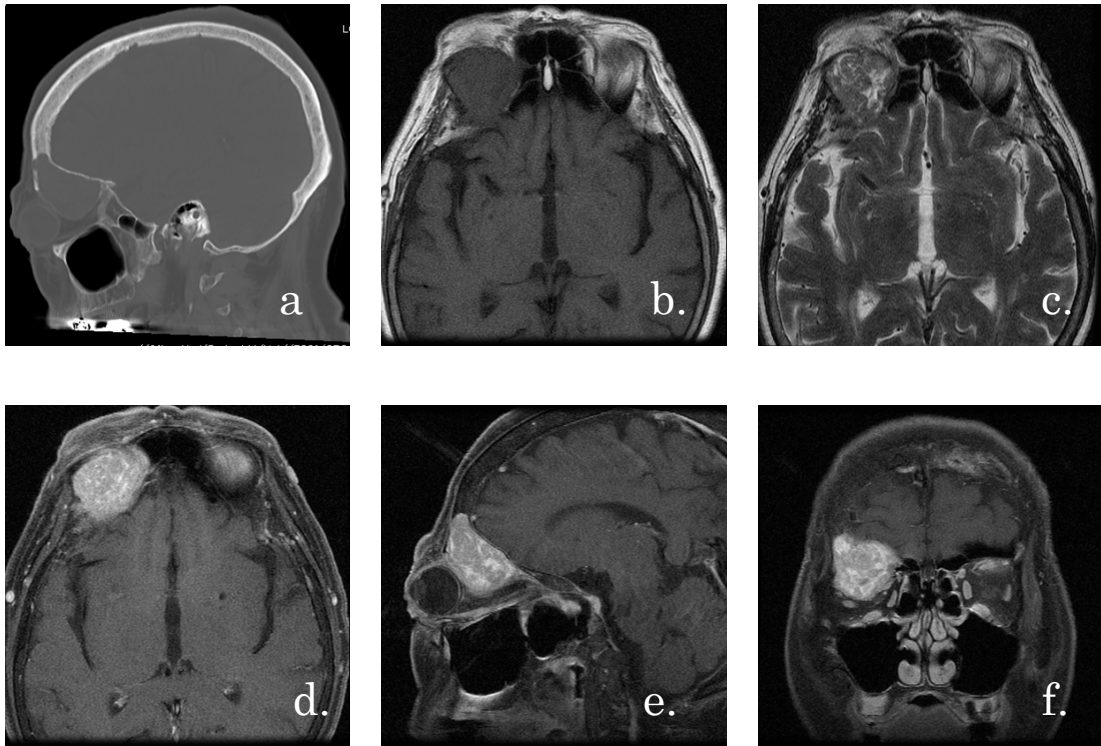


Figure 1. Preoperative images of a 71-year-old woman presenting with exophthalmos. (a).Computed tomography showing erosion of the frontal bone surrounding the right frontal sinus. (b), (c). Cranial magnetic resonance imaging demonstrated a right frontal sinus tumor isointense on T1- (b) and mixed iso- and hyper-intense on T2-weighted images (c). (d), (e), (f). The tumor was heterogeneously enhanced and compressed the right orbit.

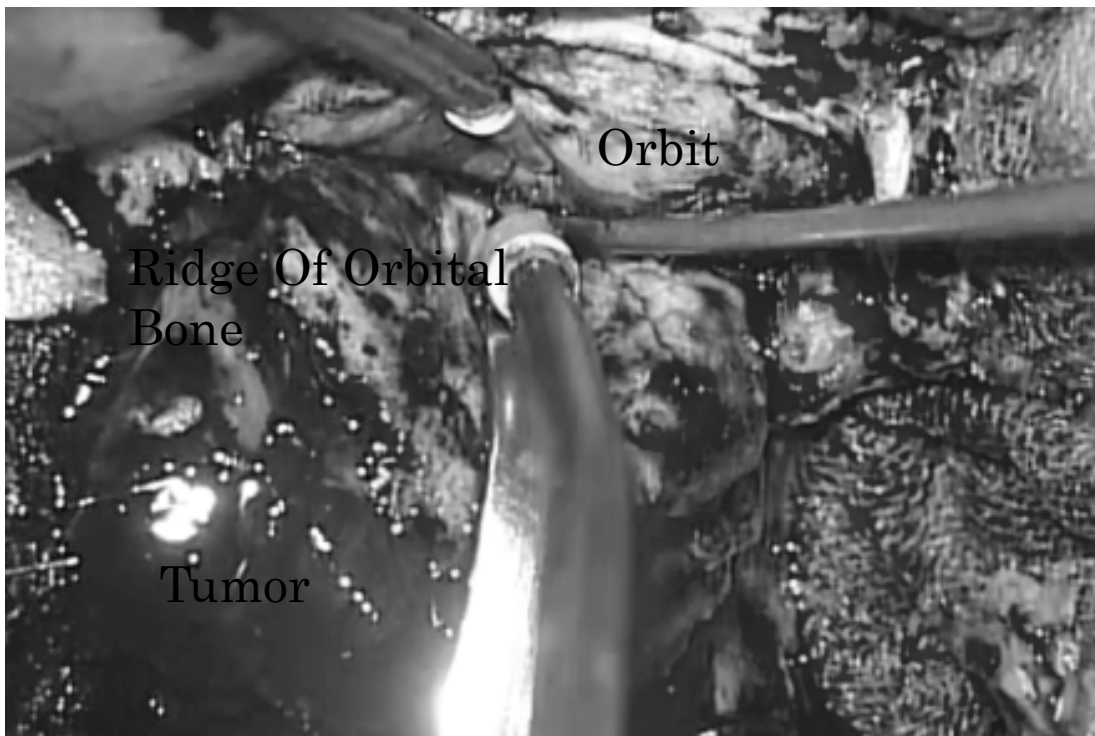


Figure 2. Intraoperative exposure showed a well circumscribed tumor without invasion into the brain or orbit.

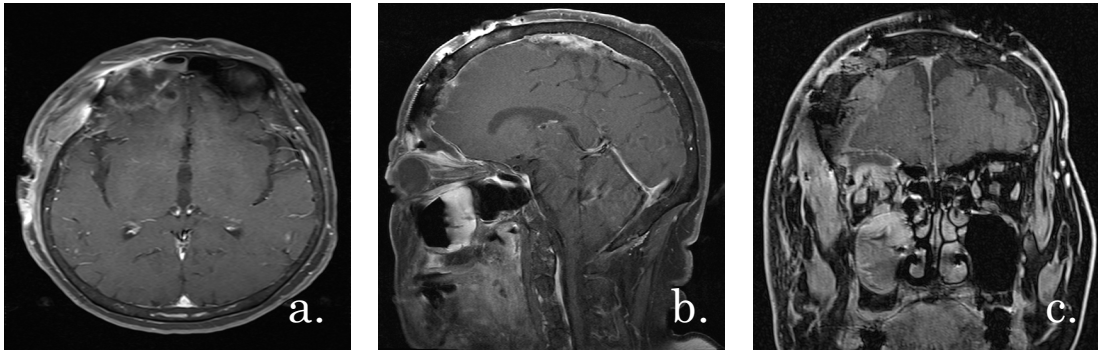


Figure 3. Postoperative cranial MRI confirmed total resection of the sinonasal tumor.

from BML, the clinical course of these tumors is typically inactive.^{1,11}

A definitive diagnosis of BML should only be made after careful review of the previous uterine leiomyoma and exclusion of a missed malignancy. The most common site of metastasis is the lung¹⁴; although cases with involvement of the lymph nodes, heart, skull, spine, and retroperitoneum have been reported.⁹ In our patient, meticulous histologic review of the resected uterine tumor confirmed our diagnosis of BML; the features of the sinonasal tumor was similar to that of the original tumor. No other tumors were detected on CT scans of the lung and abdomen obtained before the second operation. To our knowledge ours is the first reported case of BML to the nasal

sinus.

The etiology of BML continues to be debated. Cho *et al.*⁶ suggested low-grade sarcoma with metastatic potential as the primary lesion and attributed its deceptive benign appearance to sampling error. Others posited hematogenous spread of a benign uterine leiomyoma^{1,12}, because a significant number of women with BML had undergone dilatation and curettage, hysterectomy, or myomectomy that may have led to the vascular spread of uterine leiomyoma cells.

The optimal therapy for BML has not yet been established and the first treatment choice is surgical resection of the tumor.⁴ In patients with unresectable BML, hormone treatment is an alternative choice of therapy.¹³

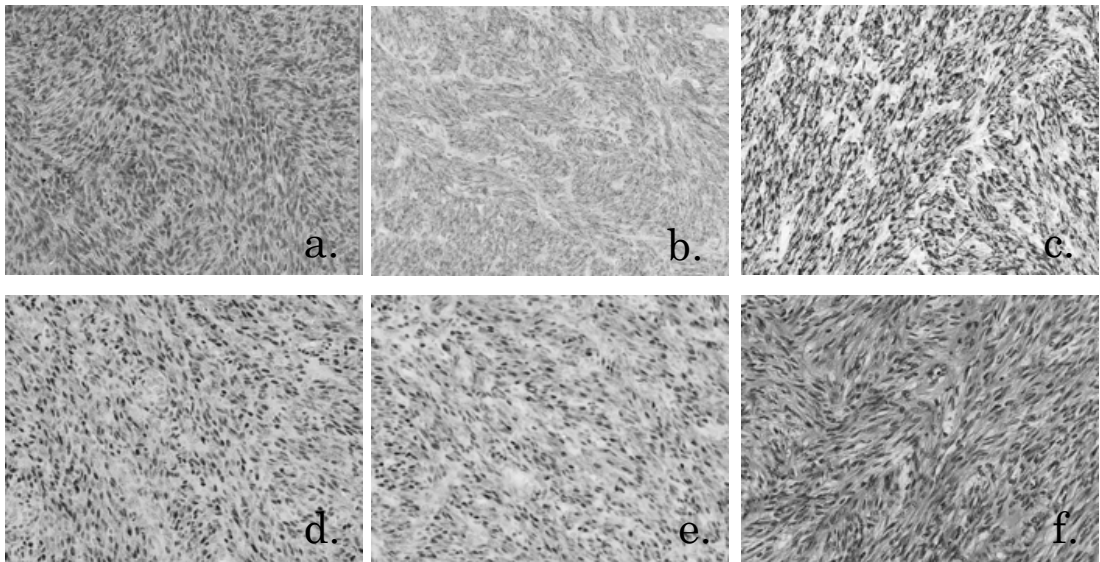


Figure 4. (a). Hematoxylin and eosin staining showed spindle-shaped smooth muscle cells with eosinophilic cytoplasm proliferating in an anastomosing fascicular pattern. No malignant features were observed. (b), (d), (e). The tumor cells were diffusely positive for α -smooth muscle actin (b) and desmin (c). The nuclei stained positive for estrogen receptor (d) and progesterone receptor (e). (f). The uterine leiomyoma resected four years earlier exhibited the same histologic features (H&E). All magnifications x 200

In conclusion, we report a rare sinonasal leiomyoma whose histologic features was similar to a previously resected uterine leiomyoma. To our knowledge this is the first report of a benign metastasizing leiomyoma to the frontal sinus.

DISCLOSURE

Conflict of interest: None

REFERENCES

1. Abramson S, Gilkeson RC, Goldstein JD, Woodard PK, Eisenberg R, Abramson N. Benign metastasizing leiomyoma: Clinical, imaging, and pathologic correlation. *AJR Am J Roentgenol* 2001; 176:1409-13.
2. Agarwal AK, Bansal R, Singhal D: Sinonasal leiomyoma. Report of 2 cases. *Ear Nose Throat J* 2005; 84:224, 226-30.
3. Ardekian L, Samet N, Talmi YP, Roth Y, Bendet E, Kronenberg J: Vascular leiomyoma of the nasal septum. *Otolaryngol Head Neck Surg* 1996; 114:798-800.
4. Arif S, Ganesan R, Spooner D. Intravascular leiomyomatosis and benign metastasizing leiomyoma: An unusual case. *Int J Gynecol Cancer* 2006; 16:1448-50.
5. Bloom DC, Finley JC Jr, Broberg TG, Cueva RA. Leiomyoma of the nasal septum. *Rhinology* 2001; 39:233-5.
6. Cho KR, Woodruff JD, Epstein JI. Leiomyoma of the uterus with multiple extrauterine smooth muscle tumors: A case report suggesting multifocal origin. *Hum Pathol* 1989; 20:80-3.
7. Farah-Klibi F, El Amine O, Ferjaoui M, Ben Jilani S, Zermani R. Sinonasal leiomyoma: An exceptional localization. *Tunis Med* 2008; 86:752-4.
8. Harcourt JP, Gallimore AP: Leiomyoma of the paranasal sinuses. *J Laryngol Otol* 1993; 107: 740-1.
9. Jo JH, Lee JH, Kim DC, *et al.* A case of benign metastasizing leiomyoma with multiple metastasis to the soft tissue, skeletal muscle, lung and breast. *Korean J Intern Med* 2006; 21: 199-201.
10. McCaffrey TV, McDonald TJ, Unni KK: Leiomyoma of the nasal cavity. *Laryngol Otol* 1978; 92:817-9.
11. Rakhshani N, Hormazdi M, Abolhasani M, Shahzadi M. Benign metastasizing leiomyoma of the uterus. *Arch Iran Med* 2007; 10:97-9.
12. Rao AV, Wilson J, Sylvester K. Pulmonary benign metastasizing leiomyoma following hysterectomy: A clinicopathologic correlation. *J Thorac Onco*: 2008; 3:674-6.
13. Rivera JA, Christopoulos S, Small D, Trifiro M. Hormonal manipulation of benign metastasizing leiomyomas: Report of two cases and review of the literature. *J Clin Endocrinol Metab* 2004; 89:3183-8.
14. Sapmaz F, Ergin M, Katrancioğlu O, Gonlugur T, Gonlugur U, Elagoz S. Benign metastasizing leiomyoma. *Lung* 2008; 186:271-3.
15. Steiner PE. Metastasizing fibroleiomyoma of the uterus: A report of a case and review of the literature. *Am J Pathol* 1939; 15: 89-110.
16. Yang BT, Wang ZC, Xian JF, Hao DP, Chen QH. Leiomyoma of the sinonasal cavity: CT and MRI findings. *Clin Radiol* 2009; 64:1203-9.