Soft Tissue Sarcomas of the Sella: Clinicopathological Features

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Introduction

Soft tissue sarcomas are a rare and aggressive type of sellar lesion, often associated with prior radiotherapy to the pituitary gland.

Methods

Patients diagnosed with soft tissue sarcomas of the pituitary gland at Partners Healthcare hospitals from 1999-2013 were retrospectively reviewed for patient characteristics, presenting symptoms, radiographic and pathologic features, treatment, and outcomes; as approved by the PHS IRB.

Results

9 patients with soft tissue sarcomas of the sella were identified, including 7 unclassified sarcomas, 2 rhabdomyosarcomas, and 1 malignant solitary fibrous tumor. The median age at diagnosis was 51.9 years (range: 28.3 - 85.6) and 33% of cases were females. Patients most commonly presented with headaches (67%), cranial nerve palsies (50%), and visual field defects (33%). All patients exhibited abnormally elevated prolactin levels (100%) and 67% developed hypothyroidism. All cases underwent resection, by transsphenoidal approach in 75%, with gross total resection achieved in only 50% of cases. Surgery was followed by adjuvant chemotherapy in 60% and radiotherapy in 50% (median dose 45 Gy). Symptomatic improvement was observed in 67% of patients following treatment, with exacerbation of symptoms in the remaining third. Half of patients experienced local recurrence, at a median of 0.5 yrs following treatment. In patients with clinical follow-up, 33% of cases that achieved gross total resection recurred, while 100% of cases that underwent subtotal resection recurred.

Table 1. Tumor Subtypes

Age	Median	Range
Presentation (yr)	51.9	22-80
Sex	**n**	**(%)**
Male	6	67
Tumor Type	**n**	**(%)**
Undifferentiated Sarcoma	7/9	78
Rhabdomyosarcoma	2/9	22

Table 2. Presenting Characteristics

Presenting symptoms	n	(%)
Headache	4/6	67
Visual Field Deficit	2/6	33
Cranial Nerve Palsy	2/6	33
Memory Loss	1/6	17
Pituitary Function Labs	**n**	**(%)**
Prolactin (high)	2/2	100
TSH (low)	2/3	67

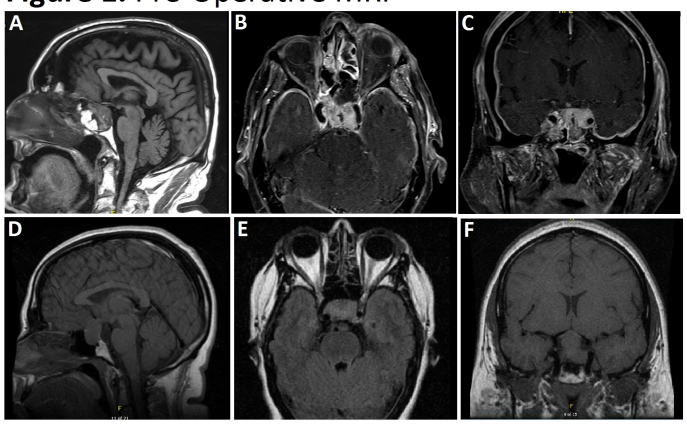
Table 4. Therapeutic Management

Surgical Approach	n	(%)
Craniotomy	1/4	25
Transsphenoidal	3/4	75
Removal Methodology	**n**	**(%)**
Microscopic	2/4	50
Endoscopic	2/4	50
Combined	0/4	0
Extent of Resection	**n**	**(%)**
Biopsy	0/4	0
Subtotal Resection	1/4	25
Gross Total Resection	3/4	75
Other Modalities Employed	**n**	**(%)**
Chemotherapy	5/7	71
Radiotherapy	4/7	57

Table 3. Histological Staining

Positive Reactivity	n	(%)
Vimentin	2/2	100
ACTH	2/3	67
Synaptophysin	2/4	50
EMA	3/7	43
Negative Reactivity	**n**	**(%)**
S100	7/7	100
AE1/AE3	2/2	100
Chromogranin	5/6	83
CD34	5/6	83
Keratin	7/9	78

Figure 1. Pre-Operative MRI



Two illustrative examples of sarcomas of the sella are shown here (A, B, C and D, E, F)

Table 5. Post-Operative Outcomes

Follow-Up	Total Available	Median	Mean	Range
Years	5	1.7	1.4	0.01-3.02
Presentation	**Improvement**	**No Change**	**Decline**	**New Symptom**
Headache	2/2	0/2	0/2	0/15
Visual Field Deficit	3/5	0/5	2/5	1/12
Cranial Nerve Palsy	6/9	0/9	3/9	1/7
Recurrence	**n**	**(%)**		
Total Recurrence	2/4	50		
Median PFS* (yr)	0.47			

*Progression-Free Survival

Conclusions

Soft tissue sarcomas of the pituitary are aggressive tumors that may mimic more benign tumors on imaging, but can locally destroy the intrasellar tissues, invade into the neighboring cavernous sinuses, compress suprasellar structures, and multiply recur despite multimodality treatment. Management of these lesions often requires maximal resection, adjuvant radiotherapy, and systemic chemotherapy depending on the histological subtype of the sarcoma.