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(REVIEW ARTICLE)

Primary intracranial myxoma mimicking a frontal parasagittal meningioma: A Case Report and Review of the Literature

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Abstract

Primary intracranial myxomas are exceptionally rare benign mesenchymal tumors, most commonly involving the skull base, whereas supratentorial frontal parasagittal extra-axial localizations remain exceedingly uncommon and may radiologically mimic meningiomas, making preoperative diagnosis particularly challenging. We report the case of a 58-year-old patient presenting with chronic fronto-orbital headaches complicated by a generalized tonic–clonic seizure. Brain magnetic resonance imaging revealed a well-circumscribed multilobulated right frontal parasagittal extra-axial lesion, hypointense on T1-weighted images, hyperintense on T2/FLAIR sequences, and showing marked heterogeneous gadolinium enhancement, initially suggestive of meningioma or another mesenchymal tumor. Given the symptomatic presentation, the patient underwent gross-total surgical resection through a right frontolateral craniotomy. Histopathological examination demonstrated a hypocellular myxoid tumor composed of spindle-shaped cells embedded within an abundant myxoid stroma, with immunohistochemical positivity for vimentin and S-100 protein, negativity for epithelial membrane antigen, and a low proliferative index (Ki-67 <1%), confirming the diagnosis of primary intracranial myxoma. Postoperative cardiac investigations excluded an underlying cardiac myxoma, supporting a primary intracranial origin. The postoperative course was uneventful, and follow-up magnetic resonance imaging showed no recurrence. Primary intracranial myxomas should be considered in the differential diagnosis of atypical extra-axial frontal lesions because radiological findings remain non-specific, and definitive diagnosis relies on histopathological and immunohistochemical confirmation. Gross-total resection remains the cornerstone of treatment and the principal prognostic factor for long-term disease control.

Keywords: Parasagittal meningioma; Myxomas; Primary intracranial myxomas

1. Introduction

Myxomas are rare benign mesenchymal tumors characterized histologically by a hypocellular proliferation embedded within an abundant myxoid stroma. [1–17]. Although myxomas most commonly arise in the heart, particularly in the left atrium [2–3], extracardiac localizations are exceptionally rare. Among these, primary intracranial myxomas constitute an exceedingly uncommon entity, with only a limited number of cases reported in the literature, most of them described as isolated case reports [2–13,15–17]. As a result, their true incidence, biological behavior, and optimal management remain poorly defined. Unlike secondary intracranial involvement related to cardiac myxomas, primary intracranial myxomas are thought to originate from residual embryonic mesenchymal tissue, particularly at the skull base sutures [4–8]. Their clinical and radiological presentation is highly variable and non-specific, often mimicking more common intracranial tumors such as meningiomas or other mesenchymal lesions. Consequently, preoperative diagnosis is challenging, and definitive identification relies almost exclusively on histopathological and immunohistochemical examination. Reported intracranial locations are heterogeneous, with a predominance of extra-axial skull base lesions

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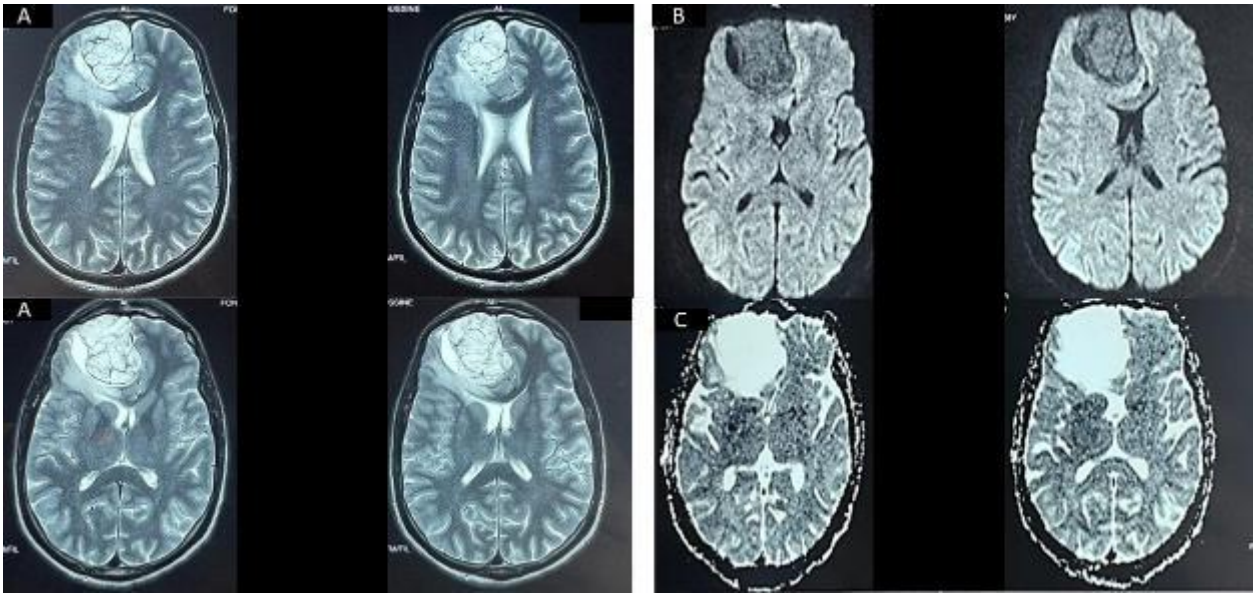
[2–13] [15–17]. Supratentorial frontal extra-axial localizations are particularly uncommon, and only a few cases have been documented to date [11]. Because of this extreme rarity, each additional well-documented case contributes valuable information to the understanding of the clinical presentation, diagnostic pitfalls, and therapeutic strategy for this unusual tumor.

In this report, we describe a rare case of a **primary intracranial myxoma presenting as a frontal parasagittal extra-axial lesion**, radiologically suggestive of a meningioma, in a 58-year-old patient. We discuss the clinical, radiological, surgical, and histopathological features of this case and review the relevant literature, with particular emphasis on diagnostic challenges and management considerations.

2. Case report

A 58-year-old patient with no significant past medical history presented with a long-standing history of chronic headaches, predominantly fronto-orbital in location, evolving over several years. Three months prior to admission, the clinical course was complicated by the occurrence of a generalized tonic–clonic seizure. There was no history of fever, and the general condition remained preserved. On admission, neurological examination revealed a fully conscious patient with a Glasgow Coma Scale score of 15. No focal motor or sensory deficits were noted, and cranial nerve examination was unremarkable.



(A) Axial T2-weighted image showing a hyperintense lesion with a thin cerebrospinal fluid (CSF) rim separating it from the surrounding cerebral parenchyma, associated with peripheral hyperintense edema.; (B, C) Diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) maps demonstrating no diffusion restriction

Figure 1 Preoperative MRI.

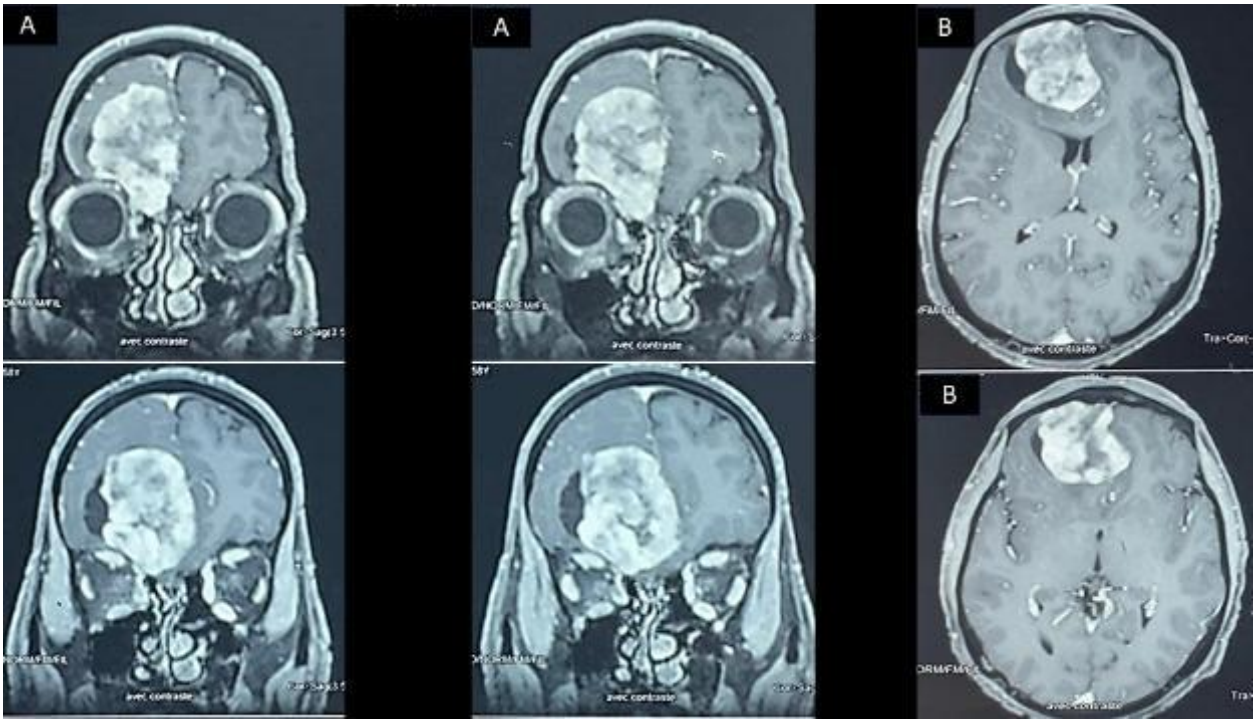


Figure 2 Preoperative coronal(A) and axial (B) MRI after gadolinium administration revealed a mass exhibiting intense contrast enhancement, with scattered central non-enhancing areas, associated with a small peripheral cystic component

Following this clinical presentation, **a brain magnetic resonance imaging (MRI) study was requested**, which demonstrated a well-circumscribed, multilobulated right parasagittal frontal space-occupying lesion. The lesion appeared hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR sequences, without diffusion restriction on diffusion-weighted imaging (b = 1000). Following gadolinium administration, the mass showed marked contrast enhancement, except for a few central non-enhancing areas, associated with a small peripheral cystic component. Surrounding vasogenic edema of the adjacent subcortical white matter was present. The lesion was located in a right frontopolar and basal extra-axial position, causing mass effect on the frontal pole and displacement of the falx cerebri, without evidence of adjacent bone involvement or dural tail enhancement. Based on these radiological features, a diagnosis of meningioma or, alternatively, a mesenchymal tumor was strongly suspected. [Figure 1-2].

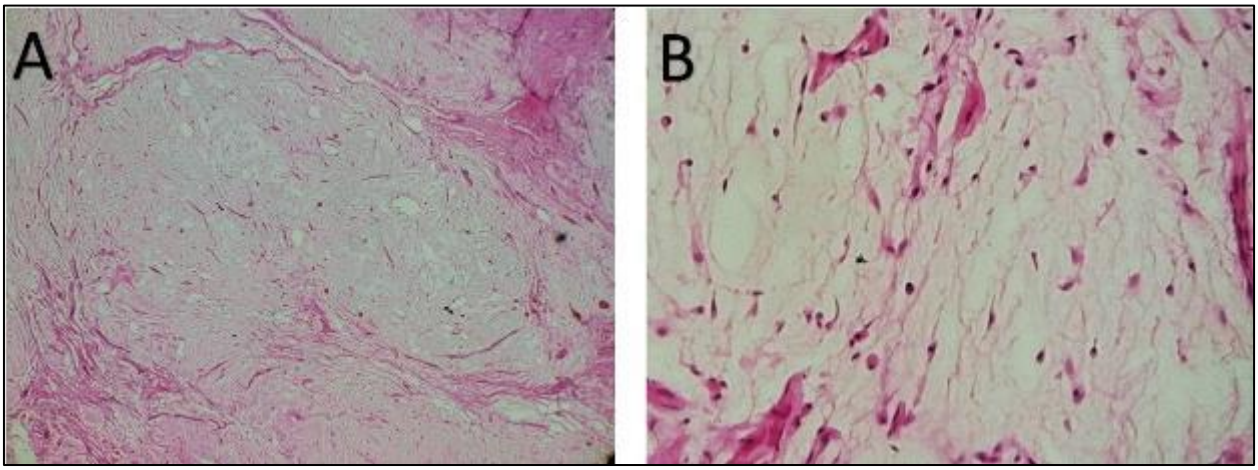
Given the symptomatic nature of the lesion, surgical resection was indicated. A complete preoperative workup was performed, including routine biological investigations and a pre-anesthetic evaluation with cardiology assessment. Transthoracic echocardiography revealed no abnormalities.

The patient was taken to the operating room and underwent surgery under general anesthesia. The head was fixed in a Mayfield head clamp, and a right frontolateral craniotomy was performed. After dural opening and dural suspension, an extra-axial mass was identified. The tumor appeared grayish-white in color, with a soft to firm consistency and low vascularity. The macroscopic appearance was initially suggestive of a meningioma. Central debulking was performed first, followed by careful dissection of the tumor capsule and its insertion base, which appeared to be falcine, in close proximity to the crista galli. A macroscopically complete resection was achieved, with coagulation of the implantation base. The surgical specimen was sent for histopathological examination.

Postoperative recovery was uneventful. The patient was extubated one hour after surgery. A postoperative CT scan performed six hours after the procedure showed no hemorrhagic complications. The patient remained in the recovery unit for 12 hours before being transferred to the neurosurgical ward.

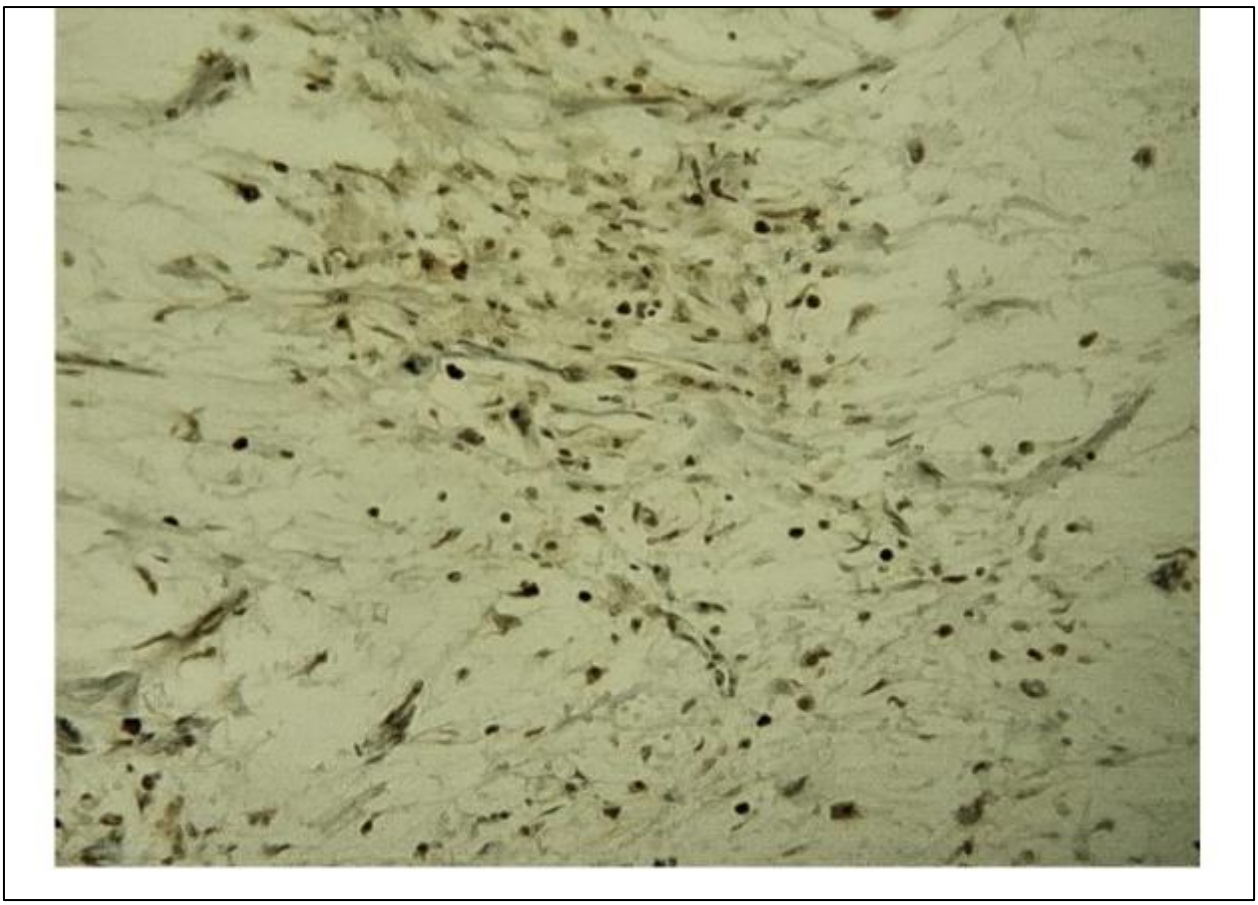
Histological examination of the surgical specimen revealed a tumor composed of spindle-shaped cells arranged in a lobulated architecture within an abundant myxoid stroma rich in mucopolysaccharides and poorly fibrillar in appearance. Tumor cells showed irregular contours with oval nuclei and vacuolated, occasionally eosinophilic cytoplasm. No mitotic figures or necrosis were identified. Immunohistochemical analysis demonstrated negativity for epithelial membrane antigen (EMA), positivity for S-100 protein, and strong positivity for vimentin. The Ki-67

proliferative index was estimated to be less than 1%. Taken together, the morphological features and immunohistochemical profile were consistent with the diagnosis of an intracranial myxoma. [Figure 3-4].



(A) Photomicrograph of a cerebral myxoma showing a low cellularity tumor proliferation with a lobulated architecture, developing within an abundant myxoid stroma (hematoxylin and eosin stain, ×10).
(B) Higher magnification showing spindle-shaped tumor cells with mild cytonuclear atypia, without mitotic figures (hematoxylin and eosin stain, ×20).

Figure 3 Histopathological findings



Immunohistochemical staining showing **tumor cell expression of S-100 protein**.

Figure 4 Immunohistochemical findings.

Following this diagnosis, additional cardiac investigations were performed during hospitalization to exclude a primary cardiac myxoma. All cardiac evaluations were normal. A final diagnosis of primary intracranial myxoma was therefore

established. A strategy of clinical and radiological follow-up was adopted. Brain MRI performed at 3 months and 1 year postoperatively showed no residual tumor or recurrence. The patient was subsequently lost to follow-up.

3. Discussion

Myxomas are benign mesenchymal tumors first described by Virchow in the nineteenth century and histologically characterized by an abundant myxoid stroma with low cellularity [1]. They occur predominantly in the heart, where they represent the most common primary tumor of the left atrium [2,3]. Neurological manifestations associated with cardiac myxomas are usually indirect, resulting from embolic phenomena or the formation of myxomatous aneurysms [2,3]. In contrast, primary intracranial myxomas represent an exceptionally rare entity. Their true incidence remains unknown, as only a limited number of cases have been reported worldwide, mostly as isolated case reports, underscoring the extreme rarity of this localization. Each additional observation therefore contributes meaningfully to a better understanding of their clinical behavior, diagnostic challenges, and optimal management.

Reported intracranial locations of primary myxomas are heterogeneous; however, most lesions are extra-axial and show a clear predilection for the skull base [4]. This distribution has been attributed to the presence of fibrocartilaginous and mesenchymal remnants along skull base sutures, particularly at the spheno-petrosal and petro-occipital junctions. Several isolated cases have described posterior fossa involvement, with lesions generally manifesting as extra-axial masses rather than true intraparenchymal tumors [5,6]. Other skull base localizations include the cerebellopontine angle, where primary myxomas may radiologically and clinically mimic more common extra-axial tumors [7]. Anterior skull base involvement has also been described, most notably in relation to the ethmoid sinus and adjacent parasinus regions, usually with an intraosseous origin [8]. Beyond the skull base, supratentorial extra-axial localizations are exceedingly rare. These include convexity lesions involving the parietal or occipital regions [9,10], as well as tumors of meningeal origin attached to the falx or dura mater [11,12]. Among these, frontal parafalcine localizations appear to be exceptionally uncommon, with only isolated cases reported in the literature [11], making the presentation observed in our patient particularly unusual. In contrast, purely intraparenchymal primary intracranial myxomas remain exceptional, and well-documented cases with unequivocal parenchymal origin are exceedingly scarce in the literature.

The pathogenesis of primary intracranial myxomas remains poorly understood. The most widely accepted hypothesis suggests an origin from residual embryonic mesenchymal cells located along fibrocartilaginous sutures of the skull base, which may undergo benign myxoid differentiation later in life [4–8]. This hypothesis is supported by the marked predilection of primary intracranial myxomas for skull base regions, including the posterior fossa, cerebellopontine angle, and anterior skull base, as consistently reported in the literature [4–8]. Beyond these classical skull base locations, alternative mechanisms have been proposed to explain rare supratentorial extra-axial presentations. These include myxoid differentiation or metaplasia of local mesenchymal connective tissue of the meninges, particularly in regions lacking identifiable embryonic remnants [9–12]. In exceptionally rare cases, a radiation-induced mechanism has been proposed, supported by isolated reports of primary intracranial myxomas developing within previously irradiated fields after a long latency period, fulfilling classical criteria for radiation-induced tumors [13]. Ionizing radiation may promote benign mesenchymal transformation through genomic instability or activation of quiescent mesenchymal progenitor cells. These primary forms must be clearly distinguished from secondary intracranial involvement related to cardiac myxomas, as well as from rare syndromic presentations associated with Carney complex [14].

Clinically, primary intracranial myxomas exhibit marked heterogeneity, largely determined by tumor location. They may occur at any age but are more frequently reported in young to middle-aged adults, whereas pediatric cases remain exceptional [13]. No consistent sex predominance has been identified. Most cases arise sporadically, while association with Carney complex remains rare [14].

Clinical presentation varies widely according to tumor topography. Skull base lesions commonly present with cranial nerve dysfunction, reflecting involvement of adjacent neurovascular structures. Posterior fossa tumors may cause headaches, vertigo, gait instability, or cerebellar signs. Intraosseous lesions, particularly those involving the temporal bone or lateral skull base, may manifest with otological symptoms such as hearing loss, tinnitus, otalgia, or a retroauricular mass [15,16]. Parasinus or anterior skull base lesions may present with facial pain or local swelling. [8]. Supratentorial extra-axial lesions of the convexity or parafalcine region are often revealed by seizures, headaches, focal neurological deficits, or, in some cases, by a palpable cranial mass, particularly in occipital or parietal localizations. [9,10]. In all reported cases of suspected primary intracranial myxoma, systematic cardiac evaluation is mandatory and typically negative, allowing exclusion of an underlying cardiac myxoma and secondary embolic intracranial involvement.

Paraclinical diagnosis relies primarily on morphological imaging, although no radiological feature is pathognomonic. Computed tomography usually demonstrates iso- to hypodense lesions, sometimes heterogeneous, with possible bone remodeling in intraosseous forms. MRI remains the reference modality, typically showing iso- or hypointensity on T1-weighted sequences and marked hyperintensity on T2 and FLAIR images, reflecting the abundant myxoid matrix. Contrast enhancement is variable, often moderate or heterogeneous. [4–18].

At the radiological stage, the differential diagnosis most frequently raised is meningioma, particularly for well-circumscribed extra-axial lesions with contrast enhancement, as was initially suspected in our case. Other differential diagnoses include solitary fibrous tumor/hemangiopericytoma, fibro-osseous lesions, chordoma, chondrosarcoma, and less commonly myxoid glial tumors or myxoid metastatic lesions, depending on tumor location and imaging characteristics. Advanced MRI sequences, including diffusion, spectroscopy, and perfusion, do not provide specific diagnostic clues. Consequently, imaging plays a crucial role in defining tumor location, extension, and surgical planning, but does not allow definitive diagnosis. The radiological features observed in our case fully illustrate these diagnostic limitations. In addition to cerebral imaging, Systematic cardiac evaluation, including transthoracic echocardiography remains mandatory, as the absence of a cardiac primary tumor and the presence of a solitary intracranial lesion strongly support a primary origin.

Definitive diagnosis of primary intracranial myxoma relies exclusively on histopathological examination. Histologically, these tumors are characterized by a markedly hypocellular myxoid stroma composed of scattered spindle-shaped or stellate cells embedded in a mucopolysaccharide-rich extracellular matrix, without nuclear atypia, mitotic activity, or necrosis, supporting their benign nature [4–16]. Histochemical staining typically shows strong Alcian blue positivity, confirming the myxoid composition of the stroma. Immunohistochemically, tumor cells usually express vimentin, while epithelial, glial, and myogenic markers are negative [4–16]. The proliferative index is low, with Ki-67 generally below 1–2% [8–12] . Although some morphological overlap exists with other intracranial myxoid tumors, the combination of marked hypocellularity, abundant Alcian blue–positive stroma, absence of atypia, and a compatible immunoprofile allows reliable distinction [4,11,12]. In the absence of a cardiac primary tumor, these findings confirm the diagnosis of a primary intracranial myxoma, as in our case.

Complete surgical excision represents the cornerstone of treatment for primary intracranial myxomas whenever feasible. Surgical management is primarily dictated by tumor location and its relationship with surrounding neurovascular structures. Lesions of the cranial base often pose significant technical challenges due to their deep location and proximity to critical anatomical structures and may require extended skull base approaches to achieve adequate resection margins. In contrast, supratentorial lesions are generally more amenable to complete excision. When gross-total resection cannot be safely achieved, subtotal resection may be considered to reduce the risk of neurological morbidity [4–17].

The role of adjuvant therapies remains limited and poorly defined. Conventional radiotherapy has not demonstrated consistent efficacy and is generally reserved for selected cases with unresectable residual disease or repeated recurrence. In the series by Zhang et al., adjuvant radiotherapy was administered after surgery in eight patients (including conventional radiotherapy and stereotactic modalities). Follow-up data, however, were available for only 11 of 23 patients. During follow-up, four recurrences were documented at postoperative years 3, 5, 6, and 9, with a mean time to recurrence of 5.9 years. Regarding radiotherapy, outcomes were heterogeneous: among the irradiated cases with available follow-up, recurrences were reported after X-knife and P32, whereas a Gamma-knife–treated lesion showed short-term radiological reduction with unknown long-term control [4]. Overall, these findings underscore both the technical challenge of achieving complete resection in skull base myxomas and the currently uncertain contribution of radiotherapy to durable local control.

To date, chemotherapy has shown no proven effectiveness and has no established role in the management of primary intracranial myxomas.

In the present case, gross-total resection was achieved, and no adjuvant therapy was administered, in accordance with current evidence and prevailing therapeutic principles.

The prognosis of primary intracranial myxomas remains difficult to precisely define owing to their extreme rarity and the absence of large prospective series with standardized follow-up. Nevertheless, the available literature suggests a generally favorable outcome, particularly when gross-total resection (GTR) is achieved. Local recurrence represents the principal adverse event and appears to be mainly related to incomplete surgical excision rather than malignant transformation. In the largest contemporary analysis, Jian-Cong Weng et al. reported outcomes from both an institutional cohort and a comprehensive review of the literature. In their institutional series, recurrence occurred in

approximately 24% of patients after a long mean follow-up exceeding seven years, with no disease-related mortality. Their pooled analysis of previously published cases confirmed the absence of reported deaths and demonstrated actuarial progression-free survival (PFS) rates of 93.0%, 80.6%, and 67.9% at 1, 5, and 10 years, respectively. Importantly, multivariate analysis identified gross-total resection as the sole independent prognostic factor significantly associated with improved PFS, underscoring the critical role of complete excision in long-term tumor control. While isolated deaths have been reported in earlier series, particularly in complex skull base lesions, more recent data suggest an excellent vital prognosis overall [17].

Overall, primary intracranial myxomas should be regarded as benign but potentially locally recurrent lesions, with long-term outcome largely determined by the extent of surgical resection. Malignant degeneration has not been convincingly documented, and prognostic assessment remains limited by the small number of reported cases and the heterogeneity of follow-up durations across published series.

4. Conclusion

Primary intracranial myxomas are exceptionally rare tumors with non-specific clinical and radiological features, making preoperative diagnosis challenging. Histopathological examination remains the cornerstone of diagnosis and allows exclusion of more aggressive myxoid lesions or secondary intracranial involvement from cardiac myxomas. Complete surgical excision represents the treatment of choice and the most reliable prognostic factor. Given the risk of delayed recurrence, prolonged postoperative follow-up is essential. Reporting additional well-documented cases is crucial to improve understanding of the biological behavior, prognostic factors, and optimal management of this rare entity.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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