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Leiomyosarcoma metastasis tumor in calvaria: A rare case report and review of the literature

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***Abstract*---**The incidence of a metastatic tumor of the calvaria is uncommon, but it has a considerable impact in the field of neurosurgery. The hematogenous metastatic spread was common in leiomyosarcoma. Calvaria is one of the most uncommon metastatic locations for this type of cancer. A 70-year-old man with a history of rhabdomyosarcoma and thyroid cancer came with a parietal mass that rapidly enlarged, followed by sudden hemiparesis and severe cognitive impairment. Head CT scan revealed a destructive soft tissue mass, indicating a metastatic development and mass effect of tumor pushing the brain parenchymal with slight compression of the right lateral ventricle and midline deviation of 8 mm to the left side. Perioperative embolization was done, and the tumor was effectively excised through extensive excision. Histopathology indicated a malignant spindle cell mesenchymal tumor, indicating leiomyosarcoma. After the operation, the patient's neurological status was stable, and there was no sign of tumor recurrence. Recommendation for managing metastatic calvaria tumor based on histopathological findings is insufficient. The information in this case report is essential for scholarly purposes and the participant gave written informed consent for publication. For the treatment of metastases in the skull, complete excision of osteolytic calvaria lesions is indicated. Perioperative embolization can aid in the removal of a calvaria tumor with significant vascularity.

***Keywords*---**Metastatic Skull Tumor, Calvaria Tumor, Leiomyosarcoma.

Introduction

Oncological advances have enhanced the life expectancy of individuals with malignancies, particularly those that can metastasize, raising the risk of consequences at the site of metastasis (Jones et al., 2021). Calvaria metastases can be identified clinically, although they are most commonly discovered at autopsies or on magnetic resonance imaging (MRI) during the cancer staging process (Mitsuya et al., 2011).

Although calvarial metastatic lesions are less symptomatic than those in the brain parenchyma, they can become uncontrolled and compress the dural sinuses and cranial nerves. Metastatic tumors of the calvaria are uncommon however, they do happen frequently in the field of neurosurgery. Calvarial metastatic cancers account for 1-4 percent of all malignancies in bone (Wang et al., 2019). The skull, like other bones, is a common site for cancer metastases (Selbi et al., 2019).

The smooth muscle of the uterus, the digestive tract, and the walls of blood vessels are the most prevalent sites for leiomyosarcomas. They spread through the bloodstream, just like other soft tissue and bone malignancies. The most prevalent sites for metastases are the lungs, liver, and peritoneal surfaces (Barbetakis et al., 2009). However, in a small number of cases, these tumors can spread to the central nervous system, soft tissues, bones, and kidneys (Bartosch et al., 2017).

Skull metastases have gotten a lot less attention in the neuro-oncological area, which means there aren't many clinical trials comparing current therapy to evidence-based practice guidelines. This case report should perhaps contribute to the development of procedures for treating cases with skull metastases.

Method

The patients involved in this case report were patients who were hospitalized and underwent surgery at the Dr. Soetomo General Academic Hospital in Surabaya Indonesia. The patient underwent preoperative examination, investigation, and postoperative examination at the same hospital. Primary data was obtained from direct history taking to the patient and other secondary data came from medical records which contained information on supporting laboratories and supporting radiology. The patient has agreed and signed the informed consent that it will be used as an interesting case to be published for humanitarian purposes and the advancement of science. All patient identities are not disclosed in the publication to maintain patient privacy.

Discussion

Case Description

A 70-year-old male complained of a lump on the head, enlarging rapidly for 7 months before admission. The mass was first felt 2 years ago, but small in size. The pain was felt upon palpation. The patient also complained of left hemiparesis

10 days before admission. The weakness occurred suddenly after waking up. The patient also had left facial palsy. Following acute hemiparesis, the patient was admitted to the regional hospital for 3 days. In the last 10 days, the patient had a progressive deficit in memory.

Four years ago, the patient was diagnosed with rhabdomyosarcoma of the left bicep, At that time complaints, a growing lump appeared on the left upper arm. The patient then underwent surgery to remove the tumor and some of the muscle. PA results: Rhabdomyosarcoma grade III, and have been treated with radiotherapy 36 cycles. Clinical photos of the history of tumor resection in the right upper arm can be seen in Figure 1.

Nine months before admission, the patient was also diagnosed with papillary thyroid carcinoma and had undergone thyroid resection. Thyroidectomy was performed with pathological results of Papillary Carcinoma Follicular Variant, limited to the thyroid, metastases to parathyroid lymph nodes (-), and angioinvasion (-). He routinely consumed levothyroxine 100 mg once a day for 7 months.

His physical examination revealed a diminished motor strength (4/5) of the left extremities. On local examination, there was a massive lump the size of 12x8x5 cm. The color of the lump is the same as the skin. Upon palpation, the lump adhered to the bone with a soft consistency. The pain was felt by the patient upon palpation (Figure 2).

The radiological examination was done with a CT scan, revealing destruction of right parietal calvaria with bulging soft tissue mass. The mass sized 7.1 x 13.2 x 8.6 cm. There's a mass extension to the adjacent epidural region, pushing the brain parenchymal with slight compression of the right lateral ventricle and midline deviation of 8 mm to the left side. Chronic lacunar infarct was seen on right pons, right putamen, left caudate nucleus and left thalamus. These findings were consistent with the metastatic process (Figure 3).

As there is concern regarding high vascularization of the tumor, transfemoral cerebral angiography (TFCA) was done. Angiogram of the right Carotid Communis Artery (CCA) revealed a blush tumor originating from the dominant feeder of the right external carotid artery (70%), accessory middle meningeal artery (MMA), MMA, branches of the superior temporal artery (STA), and the occipital artery. A minimal feeder is seen from the pial (<30%) providing vascularity to the capsule. An Angiogram of the left CCA revealed minimal blush of tumor originating from the contralateral middle meningeal artery. An Angiogram of the right vertebral artery (Lt VA) revealed no feeding artery to the tumor. Pre-operative embolization was done on STA, accessory MMA, and MMA using a 1.9 Fr microcatheter and 200 PVA particles. The effectiveness of embolization is 60-70% of the total feeding artery (Figure 4.). Surgical resection was planned 2 days after the pre-operative embolization. The surgical steps were explained in detail in Figure 5.

Macroscopic examination revealed a soft tissue mass sized 12x9x7 cm, weighted 415-gram, smooth surface, with solid bone tissue on the margins (Figure 6). Microscopic examination revealed elongated tumor cells with a round nucleus

until spindle and mitosis. In the cytoplasm of eosinophils, arranged in irregular bundles, the surface of the tumor is limited by fibrous connective tissue, some of which grow invasively between the matrix of immature bone tissue. The trabecular tumor is attached to the dura mater. Angio-invasion was encountered. The edge of bone resection does not contain an invasive tumor.

One week following the surgery, a CT scan evaluation was done (figure 7), showing complete mass removal, Calvaria defect in right frontotemporoparietal postoperative acrylic osteoplasty, minimal edema, and midline shift to the left (6,6 mm). One year following the surgery, the patient had no complaint. The education given to patients when they leave the hospital includes: routine check-ups to the doctor for monitoring complaints, surgical wounds, and the patient's condition, patients are also educated to start practicing walking and consuming nutritious foods high in protein, fiber, and vitamins. Currently, the patient has no complaints, and the general condition is good. The patient can perform daily activities independently (Figure 8).

Tumors in tissues and organs tend to metastasize to bone, and calvaria is no exception. Leiomyosarcoma is a malignant tumor that develops from smooth muscle cells and may thus grow in practically any part of the body. Cases with metastases to the central nervous system and skull are pretty uncommon, which is why this case report was chosen as an ideal chance to explore these tumors' metastases. Metastases of leiomyosarcoma are common in the lungs and liver, although bone metastases are uncommon (only approximately 3% of cases have been recorded) (Bartosch et al., 2017). There have been several reports of intracranial metastasis. The occurrence of metastases to the skull (calvaria) suggests an advanced stage, which is cause for concern that the main neoplasm has progressed to a very advanced stage (Kamian et al. 2020; Maier et al., 2021; Selbi et al., 2018).

The patient in this case report, a 70-year-old man, was admitted to the hospital in September 2017 with complaints of a painless, well-defined lump on the right side of the head (right parietal region) for the last two years and weakness in the left half of the limbs in the past 10 days, accompanied by acute memory loss. He had undergone surgery and radiotherapy for rhabdomyosarcoma of the humerus and thyroidectomy due to thyroid cancer. In this patient the risk factors for metastases are quite a lot, considering the previous history of the disease. The patient had a history of rhabdomyosarcoma and thyroid cancer that appeared in the last 5 years. From the medical data, the patient has a history of grade III rhabdomyosarcoma, which means it appears in an unusual body location, namely on the upper arm, and has a size of more than 5 cm. Rhabdomyosarcoma itself is a malignant tumor that arises in striated muscle cells. Clinically, the location and the lumps that appear are consistent with the general picture of a skull tumor. So clinically, a lump seen on the head can be suspected of metastasis. MSCT examination of the head without and with contrast performed showed destruction of the right parietal bone with a bulging soft tissue mass with the extension of the mass to the epidural which forced the brain parenchyma so that there was a midline deviation of 8 mm to the left. With a picture of a supporting metastatic process. Chronic lacunar infarcts were also found in the right pons, right putamen, left caudate nucleus, and left thalamus. This supports the clinical

occurrence of left hemiparesis because of the mass effect pressure on the right pyramidal tract.

There is currently no standard or standard algorithm for the therapy of metastases in calvaria cancers based on histological findings, metastasis extent, and concomitant comorbidities (Özgiray et al., 2016). Because of the minimal operational morbidity, full excision of osteolytic calvarial lesions with bone restoration should be undertaken whenever possible. This patient received supportive medical care as well as surgical intervention in the form of craniotomy and tumor removal. Because of the invasive character of this tumor, extirpation here refers to the removal of the whole tumor mass, including its capsule and surrounding bone tissue. The last head CT scan before the surgery revealed that the tumor was eroding bone and pushing into the parenchyma. During surgery, the tumor and surrounding bone are removed, followed by the opening of the dura mater and removal of the tumor portion linked to the sinus. The existence of neurological impairments in the right pons, right putamen, left caudate nucleus, and left thalamus as a result of tumor compression in the parenchyma was one of the grounds for surgery in our patient. Surgery is advised in cases of skull metastases if there is a neurological deficiency, bone disintegration, dural infiltration, and complaints of intolerable agony, according to various publications. Another advantage of surgery is the diagnosis procedure, which includes the histological study of the tumor (Alessandrini et al., 2022; Elmaci et al., 2020).

The pattern discovered in the histological investigation led to the diagnosis of leiomyosarcoma metastases in this patient. The presence of cytoplasmic microfilament clusters in tumor cells strongly suggests smooth muscle development, which leads to the emergence of leiomyosarcoma cells. Histopathological analysis of the excised tumor tissue revealed a malignant spindle cell mesenchymal tumor, implying leiomyosarcoma. The calvarial metastasis can originate from rhabdomyosarcoma since it is a malignancy that can spread all over a patient's body (Dietel et al., 2021).

Clinical and radiological investigations are critical in making an accurate differential diagnosis (Bistazzoni et al., 2021). The diagnostic approach employed in this patient, a CT scan, is critical for detecting, localizing, and assessing the spread of the lesion. In addition to clinical and radiographic investigations, a biopsy of tumor tissue is required to complete the diagnosis procedure. To complete the diagnostic process, this patient may be subjected to an MRI examination. MRI examinations can detect tumor infiltration into the dura mater, brain surface, or cranial nerves more accurately than other tests (Badri et al., 2018).

Regular follow-up is required in patients with a history of metastases to track the development of tumor metastases in other areas of the body. Cerebral and spinal MRIs can be conducted within a year after the first procedure (Han et al., 2017). MRI aids in determining the kind, position, and connection of tumor components to the brain parenchyma, cranial nerves, and sinus dura. Dural sinus thrombosis, a potentially fatal consequence of calvarial metastatic malignancy, can be difficult to detect. When MR Angiography (MRA) is conducted, magnetic

resonance imaging (MRI) can help in the diagnosis and indicate sinus flow. Follow-up vertebral MRI after the first four months of surgery may reveal tumor penetration into the spinal canal and adjacent regions, including the pelvic bones, in some cases of calvarial metastases. Abdominal ultrasound and a chest X-ray are also required for follow-up metastases from these malignant tumors. An abdominal ultrasound examination, as well as a chest x-ray examination, are used to follow the growth of masses in the abdominal cavity, and a chest x-ray examination is used to detect the existence of metastases to the lungs (Jones et al., 2021; Özgiray et al., 2016; Wang et al., 2019).

The histological grade, tumor size, and tumor depth all influence the prognosis of leiomyosarcoma. These factors apply to all leiomyosarcomas, regardless of whether they are uterine or non-uterine. Larger tumors with bone or neurovascular involvement, as well as higher grading, are linked to poorer outcomes. Interestingly, when compared to retroperitoneal leiomyosarcoma, extremity leiomyosarcoma had a superior prognosis (Dietel et al., 2021). The high prevalence of recurring metastases is one of the reasons for the poor prognosis in leiomyosarcoma. Many cases of metastatic skull tumors are becoming more prevalent as technology advances. This is affected by the improvement of radiological equipment and the longer life expectancy of cancer patients with improved treatment and quality of life for patients (Bartosch et al., 2017; Elmaci et al., 2020; Özgiray et al., 2016).

Conclusion

The key to the successful treatment of metastatic calvarial tumors is establishing the correct diagnosis and therapy. Metastatic tumors in the calvaria can be surgically removed. Signs of neurological abnormalities can occur as a result of the increased bulk in the skull, which can compress the brain parenchyma. A thorough assessment and preparation are required to ensure that surgery causes the least amount of morbidity. There is currently no standard strategy for managing cases of metastatic cancers in calvaria.

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Figure

Figure 1: Clinical Photo of Tumor Resection Scar on the Right Upper Arm. History of rhabdomyosarcoma resection surgery in 2013

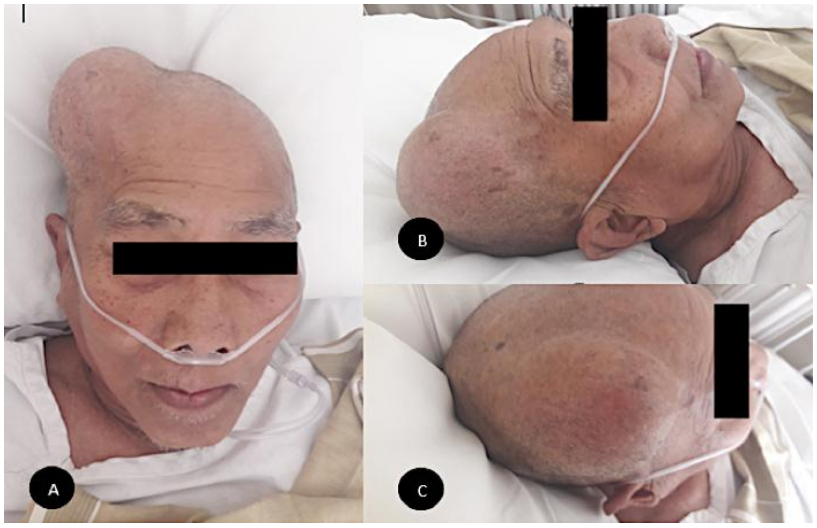

Figure 2: Clinical finding of the massive lump on the right parietal region.

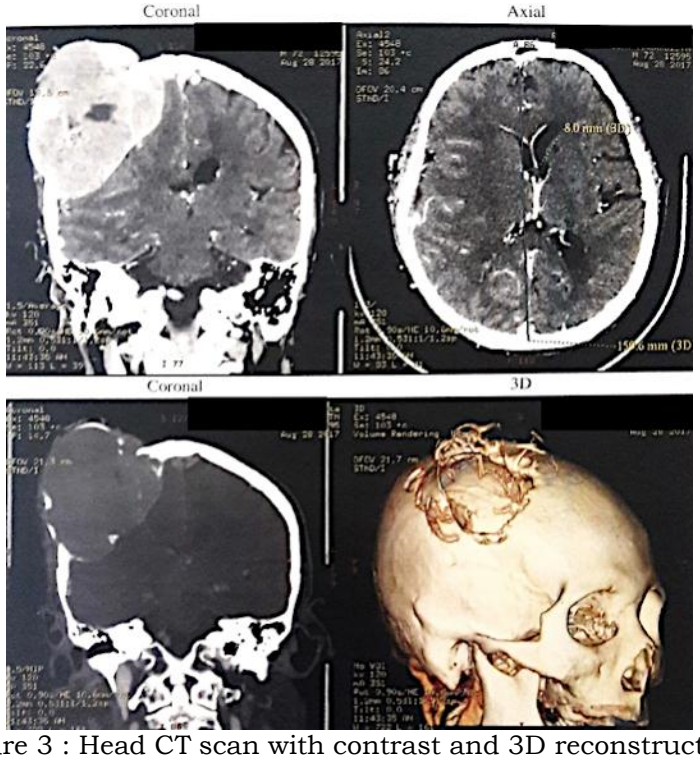

Figure 3 : Head CT scan with contrast and 3D reconstruction.

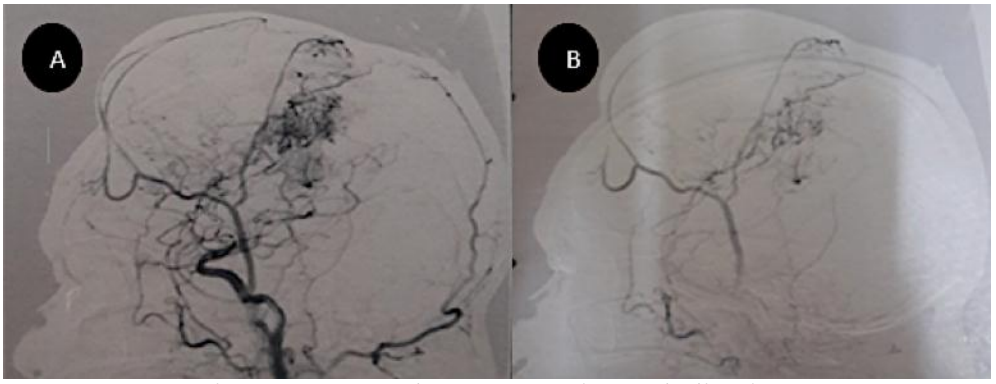

Figure 4: Pre and post-operative embolization.

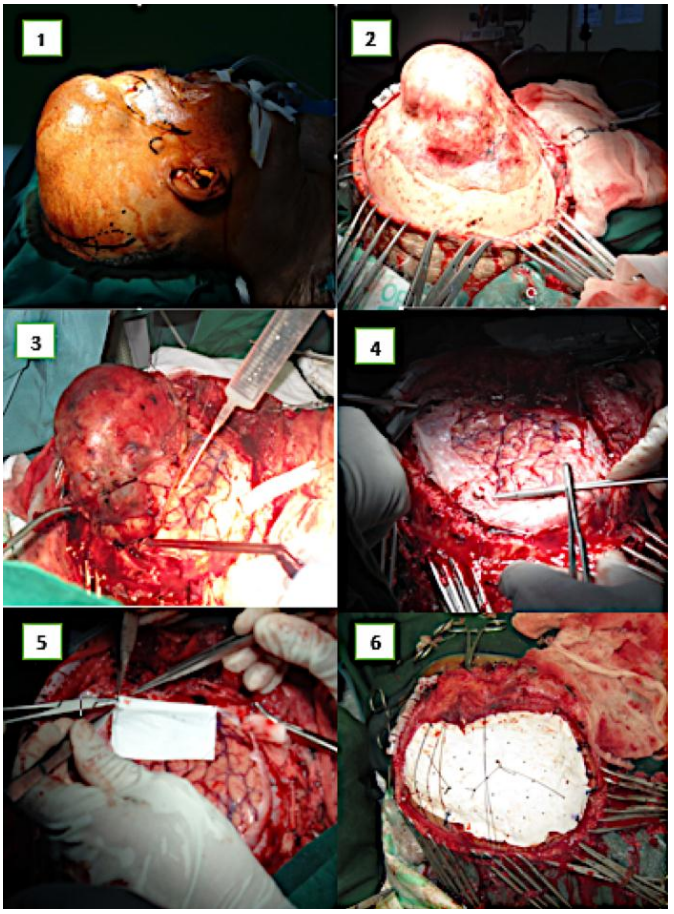

Figure 5: Operative stages. (1). Patient positioning, incision design, and disinfection; (2) removal of skin from the calvaria; (3) bone and tumor removed; (4). the dura mater is opened and the part that is attached to the sinus is removed; (5) a dural graft is placed; (6) closure layer by layer.

Figure 6: Macroscopic examination of tumor tissue

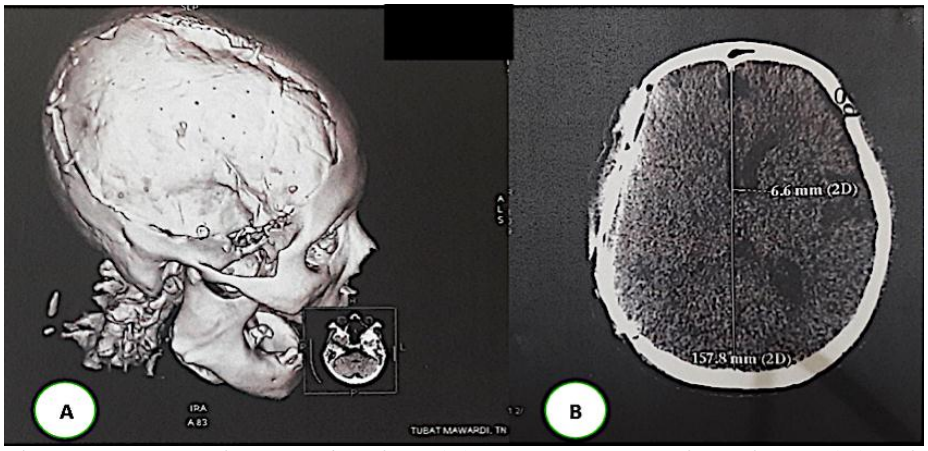

Figure 7: CT Scan Patient Evaluation. (A) 3D Reconstruction Pieces; (B) Axial cut.

Figure 8: Clinical photos 1 year following the surgery