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Surgical Research

Paraganglioma of The Skull as A Rare Presentation with Literature Review

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Keywords

Paraganglioma, Skull base tumor, Rare presentation, Paraganglioma diagnosis, Paraganglioma management.

Introduction

Paragangliomas are a rare type of neuroendocrine tumor that originates in the sympathetic and parasympathetic paraganglia [1]. The common belief is that paragangliomas are mostly benign tumors with modest growth rates, with 95% of them being non-secretory [2]. It has been discovered that this type of tumor originates embryologically from neural crest cells and can spread from the skull base to the pelvis [3].

The overall population's incidence of head and neck paragangliomas is estimated to be between 0.3 and 1 per 100,000 [1]. Head and neck paragangliomas are often non-metastatic, single tumors that are situated unilaterally [2]. In contrast to tumors of extra-adrenal origin located outside of the head and neck, which are more commonly identified in male patients [3], these tumors are typically discovered in female adults in their mid-life. The carotid body is where the majority of head and neck paragangliomas occur, accounting for up to 60% [1]. They are most typically found at the carotid artery's bifurcation within its adventitia [3]. They are followed by temporal bone paragangliomas, which account for 20-30% of all paragangliomas and are predominately female [4]. The vast majority of paragangliomas are considered benign tumors, however between 6% and 19% have been shown to have malignant tendencies and cause distant metastases [1]. The majority of metastases are identified in regional lymph nodes, however expansions to other organ systems have been discovered in 6% to 13% of patients, with the lungs and skeleton being the most prevalent targets [1]. The fundamental issue in distinguishing benign from metastatic paragangliomas is the lack of immunohistochemical criteria; the diagnosis can only be established by identifying distant metastases [5]. Paragangliomas' clinical presentation is typically determined by tumor localization [6]. Most typically, they appear to be an asymptomatic neck mass, but in rare situations, such as a vagal paraganglioma, which may cause palsies of the cranial nerves IX and XI, or when a middle ear paraganglioma is implicated, pulsatile tinnitus, hearing loss, and auditory fullness may be present [6]. Symptoms of a laryngeal paraganglioma may include dyspnea, hoarseness, and stridor caused by vocal fold paralysis [7].

Imaging is critical in assessing paragangliomas of the head and neck. CT and MRI are the first steps in the imaging examination of paragangliomas [8]. In situations with jugular-tympanic or jugular glomus paraganglioma, a CT scan provides a better assessment of the tumor's contact with the temporal bone, whereas an MRI provides a better description of the tumor's soft tissue involvement [8]. Paragangliomas had the highest uptake when analyzing SSTR expression, hence the Ga-DOTATE PET/CT was useful in distinguishing them from other cancers such as schwannoma, meningioma, and esthesioneuroblastoma, which had a lower uptake [9].

Case Presentation

70 years old female, not diabetic and not hypertensive. Referred to neurosurgery clinic from Oro-maxillo-facial department with left mandibular mass, referred to our side because of the incidental finding during systemic examination of painless left temporal mass. Recently diagnosed with thyroid begin nodule.

Her condition started 3 months prior to her presentation with painless gradually growing mass. On examination patient looks well fully conscious. There is left temple swelling with normal skin and hair measuring 6X6 cm. with mild tenderness on palpation, not pulsatile and no audible bruit. GCS: 15/15, normally reactive pupils on both sides. Normal tone, power and reflexes. No cranial nerves involvement. Other systems examination was unremarkable. CT Brain showed eroded left temporal bone with intracranial, extra-dural and extra-cranial extension figure (1 a,1 b and 1 c).

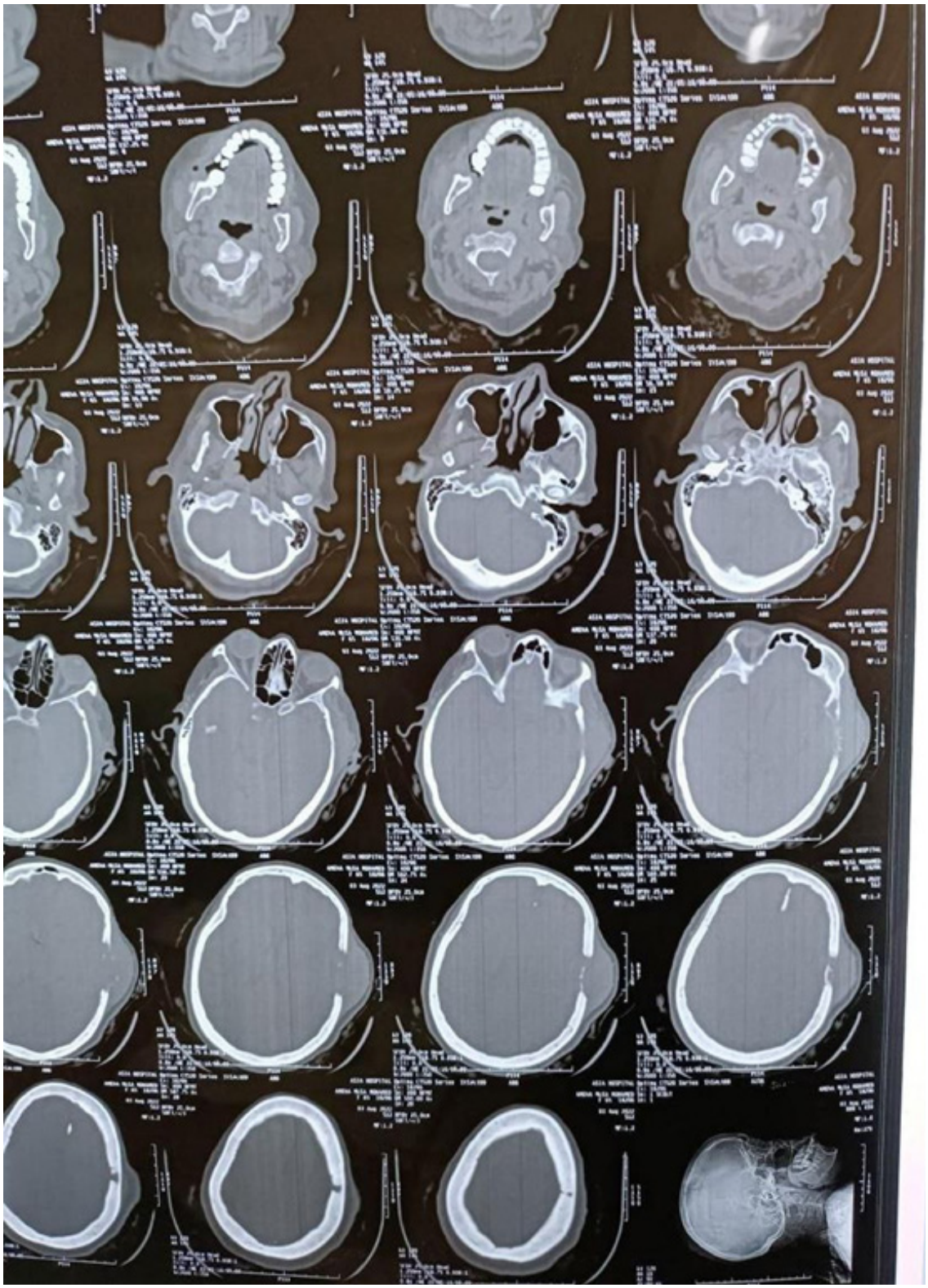

Figure 1a: CT brain, axial cut, bone window shows the bony defect caused by the lesion at the left temporo-parietal bones.

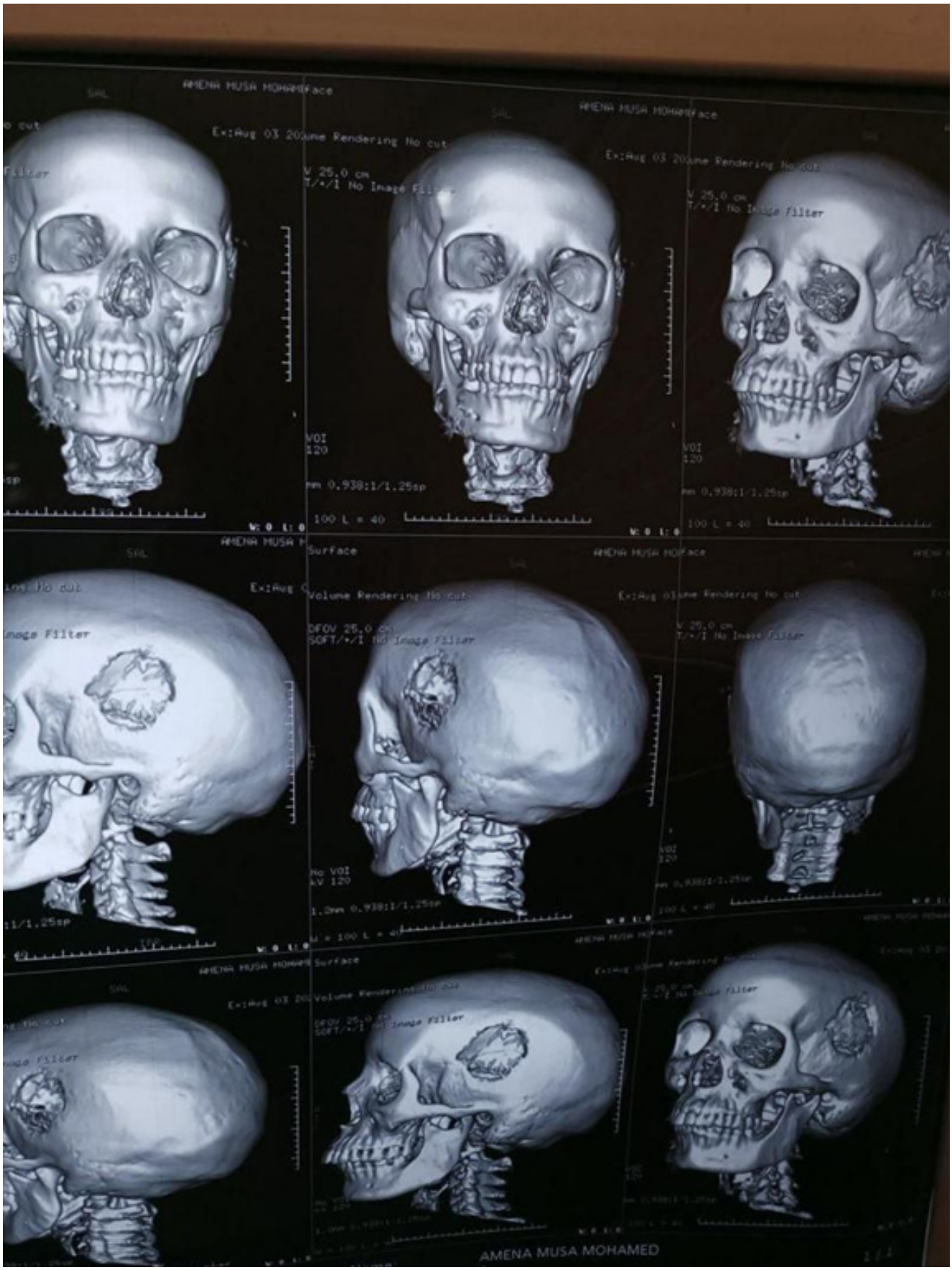

Figure 1b: 3D skull view shows the bony defect at the left temporo-parietal bones.

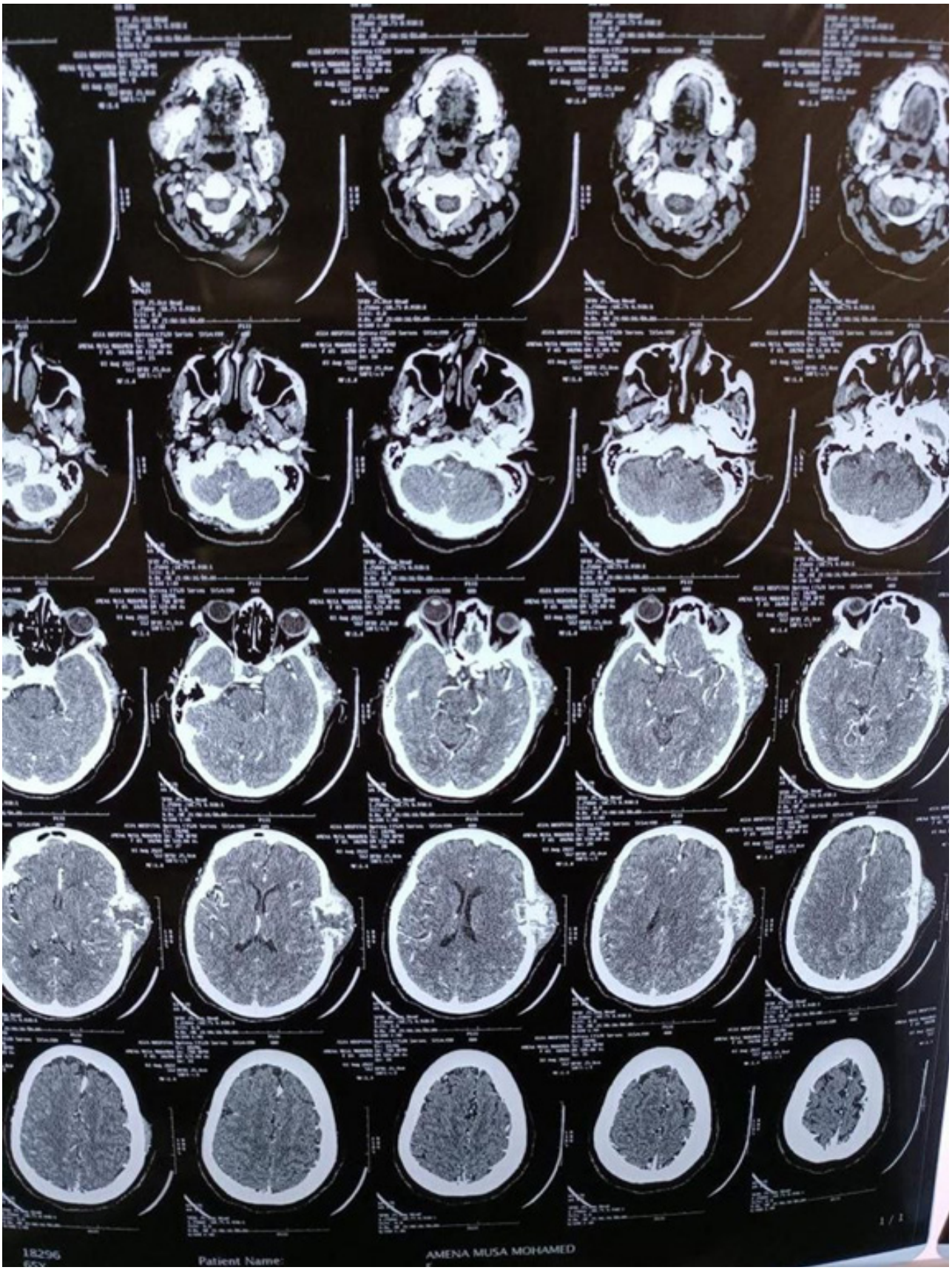

Figure 1c: CT scan with contrast, axial cut, soft tissue window shows the lesion with heterogeneous enhancement to the both intracranial and extra cranial parts.

MRI Brain with contrast showed left temporal bone lesion measuring (6.5 X 6 X 4.5 cm). With intracranial and extra cranial extension exerting significant mass effect on the left temporal lobe. This lesion shows heterogeneous enhancement after contrast administration. MRA and MRV studies showed highly vascular lesion (Figure 2a ,2b and 3).

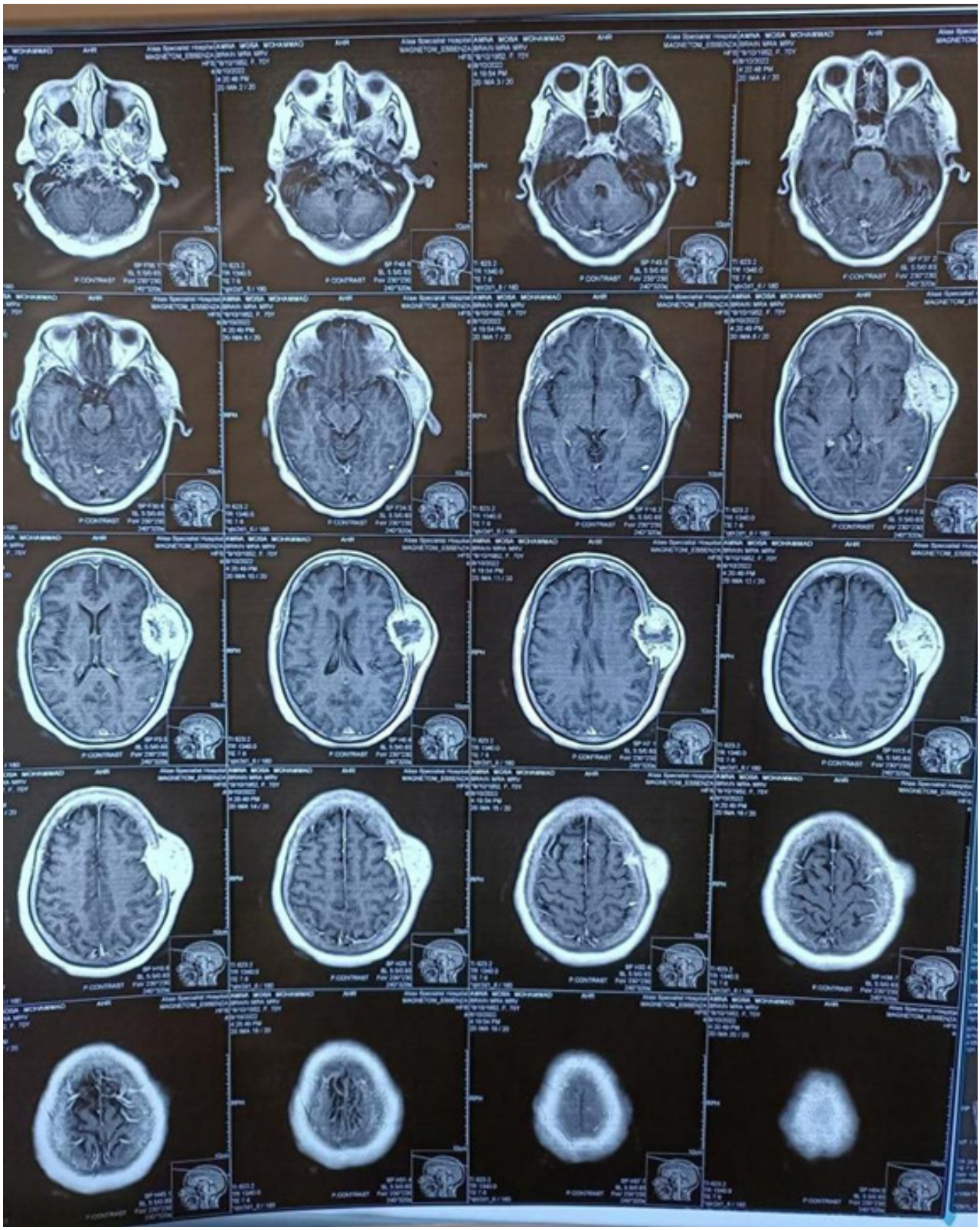

Figure 2a: MRI scan with contrast, axial cut, T1 sequence shows the intense enhancement with central hypo intensity noticed.

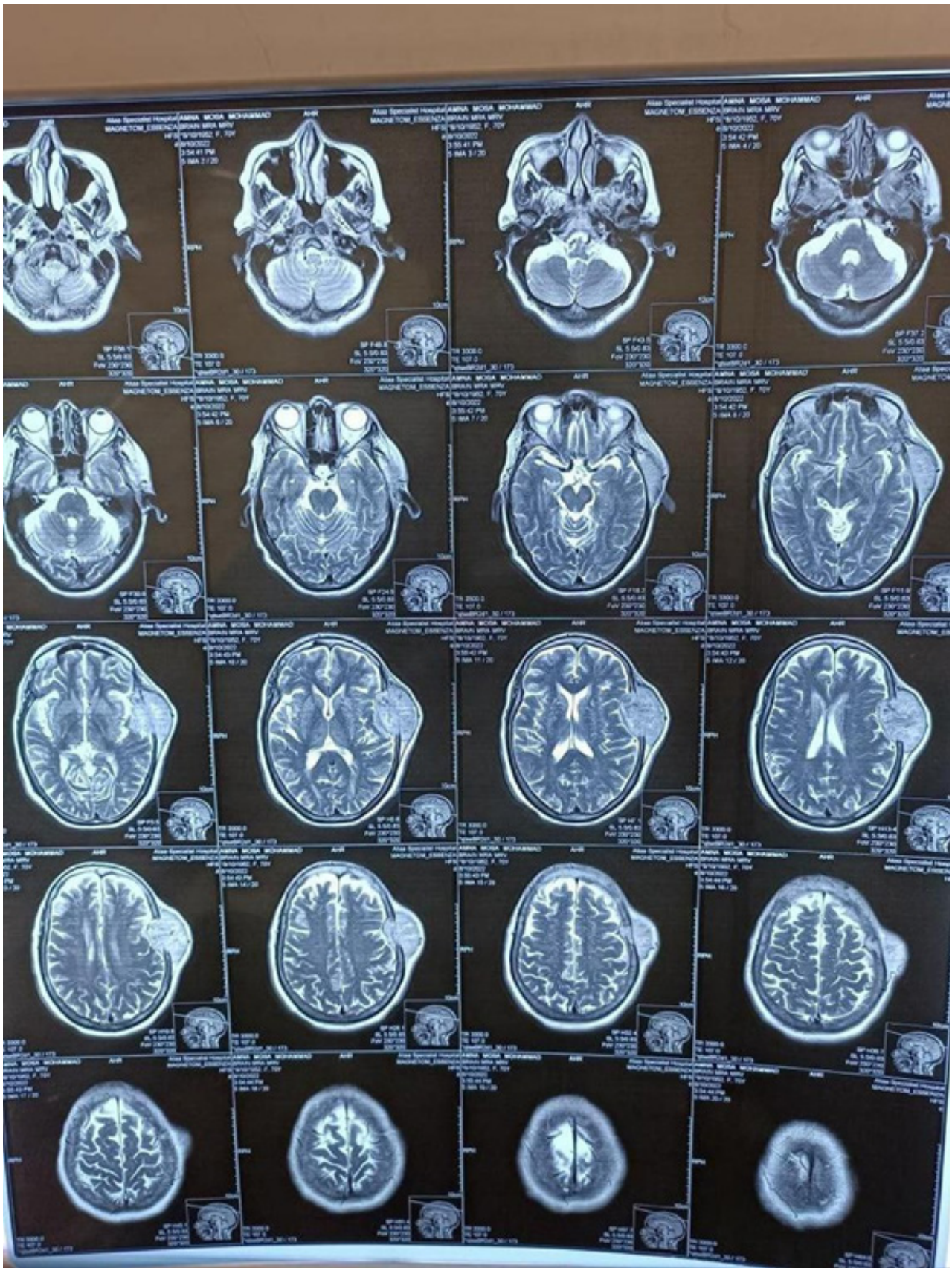

Figure 2b: MRI scan with contrast, axial cut, T2 sequence shows hyper intensity of the lesion.

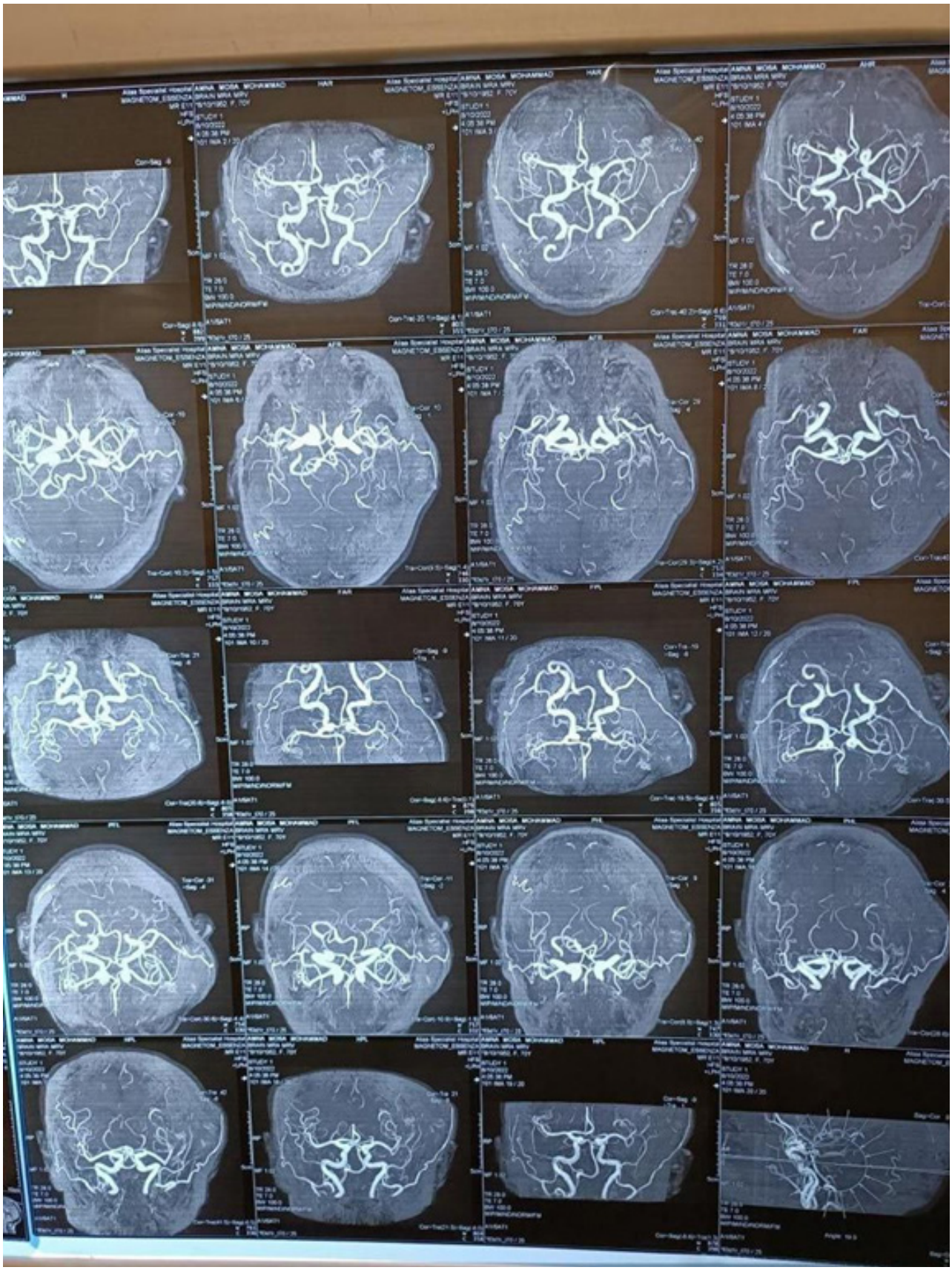

Figure 3: MRA study done for the lesion to exclude any possible vascular lesion as differential diagnosis.

Patient underwent combined surgery for the cranial lesion as well as the mandibular lesion. Gross total resection was achieved for the intracranial and the extra cranial mass. Histopathology results were suggestive of malignant paraganglioma Zell-Ballen appearance found, which was essential to diagnose paraganglioma. Immunohistochemically study confirmed the diagnosis of Paraganglioma.

Discussion

Paraganglioma has an annual incidence of 0.3 to 1 per 100,000 people, which is considered low [1]. Our case was the first to be seen in our department, and there had been no earlier reports of such cases in our country, therefore it needed to be reported. In our situation, paraganglioma was identified as enlargement of the jaw and temporal squamous bone while Paraganglioma in the head and neck is most prevalent in the carotid body (60%), temporal bone (20-30%), jugular and mastoid foramen, and vagal nerve (5%). These tumors can also develop in more specific places, including the glossopharyngeal paraganglia, jugular-tympanic PGL, larynx, nasal cavity, trachea, thyroid gland, and orbit [11].

In a comprehensive examination of this type of tumor, they discovered that head and neck paragangliomas account for a significant portion of all paragangliomas, with an estimated percentage of 65% to 70%, accounting for 0.65% of all tumors that arise at the head and neck level [1]. These tumors are typically diagnosed in female adults in their mid-life, as opposed to tumors of extra-adrenal origin located outside of the head and neck, which are more commonly found in male individuals [4]. With a prevalence of 3 to 4.1 of female patients aged 40 to 70 years [6]. Our case involved a female in her 70s. According to this review study, the prevailing assumption is that paragangliomas are, for the most part, benign tumors with modest growth rates, with 95% of them being non-secretory [2]. However, between 6% and 19% have been demonstrated to exhibit malignant behavior, resulting in distant metastases [1]. The majority of metastases are identified in regional lymph nodes, however expansions to other organ systems have been discovered in 6% to 13% of patients, with the lungs and skeleton being the most prevalent targets [1]. In our case, the main cause appears to be bone metastases. In general, temporal bone paragangliomas are the second most frequent, accounting for 20-30% of all head and neck paragangliomas [5]. Tympanomastoid paragangliomas are those that originate from Jacobson's nerve, the tympanic branch of the glossopharyngeal nerve, and Arnold's nerve, the auricular branch of the vagus nerve, whereas tympanojugular paragangliomas are those that originate from the adventitia of the jugular bulb's paraganglia [5]. Temporal bone paraganglioma tends to penetrate the skull base; the tympanojugular is positioned around the tympanic canaliculus, whilst the tympanomastoid is typically found within the jugular foramen [12]. The annual incidence of temporal bone paraganglioma has been estimated to be around 0.7 occurrences per 100,000[5]. In our instance, it primarily manifested as temporal squamous bone and jaw mets.

The fundamental issue in distinguishing benign from metastatic paragangliomas is the lack of immunohistochemical criteria; the diagnosis can only be established by identifying distant metastases [6]. These are the reasons why the World Health Organization's most recent guidelines recommend that all paragangliomas be considered malignant and that they be closely monitored even after surgical treatment [5].

Histological and immunohistochemical studies may provide valuable information for diagnosing paragangliomas and distinguishing them from other forms of cancers in the head and neck [5]. Paragangliomas of the head and neck typically have three histological characteristics: (1) the Zellballen pattern, (2) the angiomatous pattern, which consists of large spindle cells or crescent-shaped cells and capillaries, and (3) polyhedral cells with columnar architecture and abundant cytoplasm, which form the adenomatous pattern [13]. Our case histopathology microscopic features showed a partially encapsulated neuroendocrine tumor composed of nests of two populations of cells which are polygonal granular cells in zellballen pattern and peripheral sustentacular cells. The nests are separated by thin fibrous septa. some areas showed spindled sarcomatoid cells and other nests contain clear cells. Numerous vascular spaces scattered in the tumor were seen. According to the World Health Organization's most recent guidelines, an immunohistochemical examination of a specimen can reveal important details such as a high Ki67, the presence of necrosis, which is associated with other factors such as a tumor larger than 5 cm, and the presence of SDHB mutation, which may indicate malignant behavior [5]. Immunohistochemistry result of our case was with markers S100 (weakly positive), chromogranin (patchy positive), synaptophysin (patchy positive), TTF1 (negative), and Ki67 (>40), the interpretation was compatible with paraganglioma. Paragangliomas of the head and neck are typically benign tumors, with an overall 5-year survival rate of 91% for patients without distant metastases [14]. On the opposite are the patients with malignant tumors, having a 76.8% in case of lymph node metastases, and 11.8% for those with distant extensions [15].

In a similar case reported in the literature, they described a 15-year-old child with paraganglioma who had both skull and brain metastases and underwent abdomen, skull, and brain lesion excision over 5 years, with a satisfactory outcome at the 10-year follow-up. Another article published in 2011, they reported a case of paraganglioma with intracranial mets, stating that intracranial involvement is highly unusual and has only been documented rarely in the literature [16].

In 2006, an article was published concerning bone mets from head and neck paragangliomas. They described three cases of bone mets, the first of which exhibited mets to the vertebrae and occipital bone. The second case revealed mets on the petrous apex bone. The third instance reported involved metastases to the cervical vertebra. The first instance had a history of left carotid body and vagal paragangliomas with a high proliferative index on pathologic examination, whereas the third had a positive history of right carotid body tumor surgery. This is in contrast to our case, which showed a negative history for any known source, whether from the carotid body or another source, such as the belly, notwithstanding the workup we performed. In a review of

the literature from 1945 to 1995, Brewis et al [4] discovered that one-third of paraganglioma metastases were localized in bone, primarily in the vertebrae and ribs.

due to the concurrent presentation of both a jaw and a temporal lesion, it was an interesting case for both departments (maxilla-facial and neurosurgery) regarding the likehood of the pathology and so patient is scheduled for conjoint surgery and the pathology was a rare one, the malignant paraganglioma.

Conclusion

Paragnglioma may not be one of the top differential diagnosis for skull mets lesion because of its rare occurrence but should be considered especially if patient had history of paraganglioma been diagnosed elsewhere in the usual sites. immunohistochemistry always should be done when suspecting malignant paraganglioma in histopathological sample.

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