

Intracranial Lesions Presenting as Bony Swellings of Scalp – An Observational Study of 13 Cases at King George Hospital, Visakhapatnam, A Tertiary Care Government Hospital

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ABSTRACT

BACKGROUND

The purpose of this study was to evaluate the intracranial lesions eroding the calvarium and presenting as bony swellings of the scalp. This kind of presentation is usually rare. This is because intracranial tumours usually manifest early with focal neurological deficits, seizures or features of raised intracranial pressure. Only a few patients, in whom the intracranial lesions, despite being present for a long duration, do not have neurological manifestations but erode the overlying bone and may present as bony swellings of the scalp. We studied the clinical presentation of such lesions, analysed their radiological and histological characteristics.

METHODS

This is an observational study from October 2018 to September 2020. A total of thirteen cases were studied in the Department of Neurosurgery, Andhra Medical College, Visakhapatnam, Andhra Pradesh. After clinical examination, an appropriate diagnostic workup was done, and all the cases were taken up for surgery. The clinical behaviour and histopathological features of the lesions were analysed.

RESULTS

In our study of thirteen cases, the most common cause of an intracranial lesion presenting as bony scalp swelling is meningioma which comprised almost 53.8 %, the second most common lesion is secondaries. Others included fibrous dysplasia and malignant small blue round cell tumour.

CONCLUSIONS

Intracranial lesions eroding the bone and presenting as scalp swellings are rare. In our study, the spectrum of these scalp swellings has ranged from benign lesions like meningioma to malignant lesions like secondary deposits.

KEYWORDS

Bony Scalp Lesions, Intracranial Tumours with Bony Erosion, Dumbbell Brain Tumours

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BACKGROUND

The scalp is the soft tissue that covers the calvaria of the skull. The scalp extends from the external occipital protuberance and superior nuchal line posteriorly to the orbital margins anteriorly, and on the lateral aspects, it is anchored to the ears. The layers of the scalp include skin, connective tissue, galea aponeurotica, loose connective tissue and periosteum (SCALP). The lesions of scalp have a wide spectrum; they may be incidental findings in radiological studies or may present as a palpable mass over the scalp. The lesions of the scalp can be classified as congenital, traumatic, inflammatory, or neoplastic in origin. Swellings of the scalp may be first seen by a general surgeon, dermatologist, plastic surgeon, or neurosurgeon directly. Our present study aims at identifying the various intracranial lesions which present as bony scalp swellings.

Intracranial lesions rarely present as scalp swellings without having any neurological deficits, seizures or features of raised intracranial pressure. These can be primary brain tumours, secondaries from some other primary tumours or primary neoplasms of the calvarium with intracranial extensions. The presentation of intracranial tumours as swellings of the scalp is a rare clinical presentation. Primary neoplasms of the calvarium account for 0.8 % of all bone tumours of the calvarium. Benign lesions are more common than primary malignancies of calvarium.¹

Objectives

1. To study the cases that presented as a bony swelling of the scalp due to underlying intracranial lesions.
2. To note the clinical, radiological and pathological features of these intracranial lesions presenting as bony swellings of the scalp.

METHODS

This is a prospective observation study with a sample size of 13 cases conducted at Department of Neurosurgery, Andhra Medical College, King George Hospital, Visakhapatnam, Andhra Pradesh, a tertiary care Government Hospital for a period of two years from October 2018 to September 2020.

Inclusion Criteria

All intracranial lesions eroding the bone and presenting as bony scalp swellings, primary calvarial malignancies with intracranial extension are also included in the study.

Exclusion Criteria

- All the congenital swellings like encephaloceles, dermoids etc. were excluded.
- All the cases with a bony scalp swelling but without any intracranial extension were also excluded.
- Primary bone tumours of calvarium without intracranial extension were excluded.

The majority of the cases in the present study have directly presented to the neurosurgical outpatient clinic. Two cases were referred from general surgery. All these patients were evaluated radiologically with computed tomography (CT) and magnetic resonance imaging (MRI) scans. Relevant investigations were performed to determine the primary in patients where the intracranial lesion was suspected to be a secondary. Surgical workup was done for all the patients. Excised specimens were sent for histopathology for all the cases. The radiological features and the histological characteristics of tumours were analysed.

Statistical Analysis

The clinical, radiological and pathological observations were noted in each of the thirteen cases. No statistical analysis was done.

RESULTS

All the cases in the present study had presented as bony scalp swellings. None of them have any features of raised intracranial pressure, focal neurological deficits, or seizures. Radiologically the scalp can be distinguished into three layers that include the skin, the subcutaneous layer, and the galea-subgaleal-periosteal complex.² In the present study, seven cases were consistent with meningiomas which were well circumscribed and had a dural attachment and were isointense to hypointense on T1 and isointense to hyperintense on T2 weighted images with intense and homogenous enhancement on gadolinium contrast administration. The next most common lesion noted in our study was secondaries, where the lesions are typically isointense to hypointense on T1 images and typically hyperintense on T2 images with contrast enhancement, commonly with ring enhancement. In one case of fibrous dysplasia in our present study, the lesion eroded the bone. It had extended extradurally. It showed well-defined borders with sclerotic margins, and on CT there is hyperdense sclerotic lesion. On MRI, there is a heterogeneous lesion on T1 with T2 usually with contrast enhancement.

Histologically in our study, meningiomas are the most common primary intracranial tumours that presented as bony scalp swellings, which accounted for seven out of thirteen cases (53.8 %). The other common lesions are secondaries in the brain which accounted for four out of thirteen cases (30.7 %). Of these, one is from primary lung carcinoma, one from breast carcinoma, one from thyroid malignancy and one case turned out to be squamous cell carcinoma where the primary could not be identified. The other cases in the present study include malignant small blue round cell tumour and fibrous dysplasia each of which constituted one out of the thirteen cases (7.6 % each). Our study comprises a total of thirteen cases in which the most common age group affected was 41 - 50 years accounting for four of the total thirteen cases (30.7 %). The next common age group affected was 51 - 60 years and 61-70 years with each age group having an equal number of cases

that is three of thirteen cases (23.07 %). Our study had seven males (53.8 %) and six females (46.1 %).

DISCUSSION

Age of the patient, symptoms and signs, history, and meticulous clinical examination followed by necessary radiological investigations are significant factors in the preoperative diagnosis of bony swellings of the scalp, post-operative histopathological examination including immunohistochemistry when required help in the proper diagnosis and appropriate management of such cases.

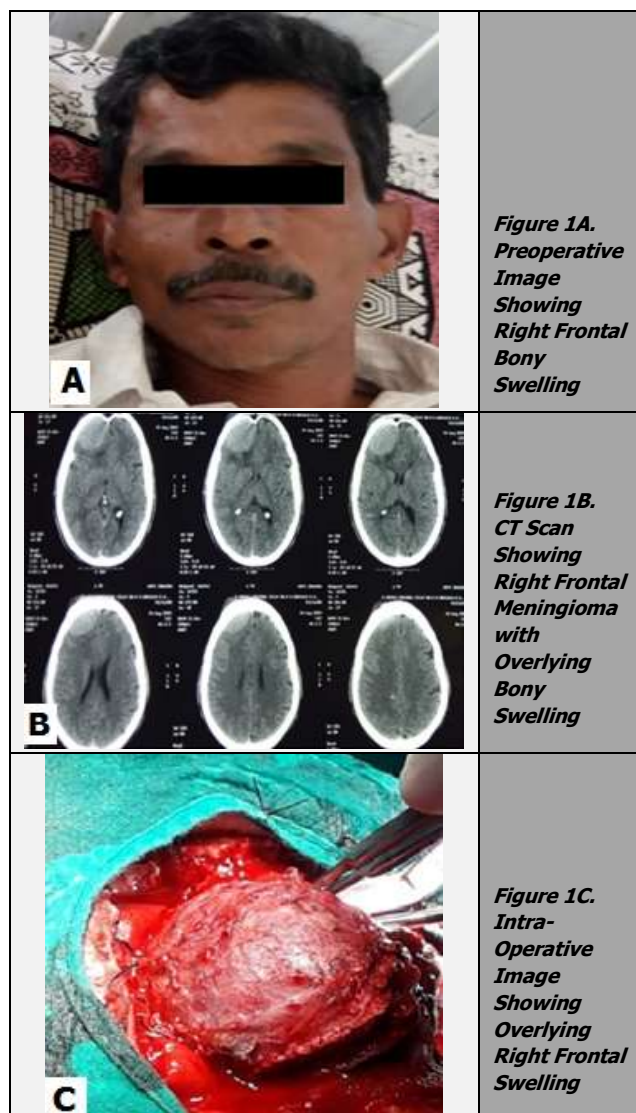


Figure 1A.
Preoperative
Image
Showing
Right Frontal
Bony
Swelling

Figure 1B.
CT Scan
Showing
Right Frontal
Meningioma
with
Overlying
Bony
Swelling

Figure 1C.
Intra-
Operative
Image
Showing
Overlying
Right Frontal
Swelling

Meningiomas (Figure 1)

Meningiomas are the most common benign intracranial tumours, and their cell of origin is arachnoid cells. They constitute roughly 13 - 26 % of intracranial tumours. A historical account of meningiomas and their surgical management highlights that "meningiomas have left their mark, in the form of hyperostosis, on human skulls as far back as prehistoric times".

In our study, we had recorded seven cases of meningiomas which eroded the overlying bone and presented with bony scalp swelling. In literature, there have been studies where the patients presented as calvarial masses, which subsequently turned out to be meningiomas.³ One of the well-known findings of meningiomas is hyperostosis of the adjacent calvarium. This hyperostosis may be a simple sclerotic change in the bone without the evidence of thickening, or it may lead to significant bone thickening and irregularity that can cause significant bony scalp swelling.

The patient below is a 47 years old man who presented to us with a painless bony swelling in the right frontal area with no other complaints. Examination revealed a swelling of bony hard consistency with no other significant neurological deficits. The patient was operated on, and the excised specimen was sent for histopathological examination, based on which it was diagnosed to be a meningothelial meningioma.

Some of the convexity meningiomas are diagnosed solely by the presence of skull or scalp swellings due to an often-associated hyperostosis and rarely due to osteolysis and tumour involving the calvarium and scalp. Our present study aims in identifying such intracranial lesions, which present as bony swellings of the scalp.

Intraosseous Meningioma

Primary intraosseous meningioma⁴ of the skull is a very uncommon lesion. It can be a differential diagnosis for the bony tumours of the calvarium. Intraosseous meningioma is a rare subtype of meningioma, accounting for less than 2 percent of meningiomas. In our present study, one case was diagnosed as intraosseous meningioma. Meningiomas that present as scalp masses and osteolytic skull lesions are considered to be aggressive in nature.⁴ Its proposed origin is the trapping of the arachnoid cells in a prior skull fracture or skull osteotomy or the embryonic remnants of the arachnoid cells with the developing calvaria^{5,6} which explains its predisposed sites near the sutures, particularly near the coronal sutures. It is predominantly seen in women in the fifth and sixth decades of life and usually revealed by a painless and expanded swelling.

Cerebral and Calvarial Metastasis⁷

Skull metastasis are malignant bone tumours, and their incidence is on the rise. The most common malignancies presenting as skull metastasis include lung, breast, prostate, malignant lymphomas etc. The patients who present with such metastases are in an advanced stage of malignancy; most of them will be either asymptomatic or may present with a painless swelling of the scalp. Metastases can have an osteolytic, osteosclerotic, or mixed pattern. Lung cancer solely is contributing to approximately 50 percent of brain metastasis in adults. In the paediatric age group, neuroblastoma is the most common cause of calvarial metastatic deposits. Calvarial metastases are not a definite clinical entity; they have specific therapeutic considerations, as suggested by a study done by Constans et al.⁸

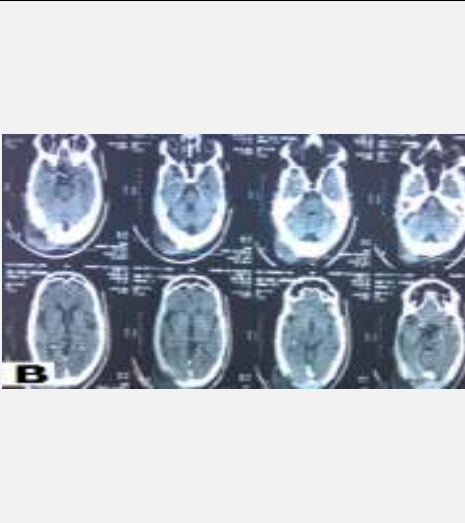
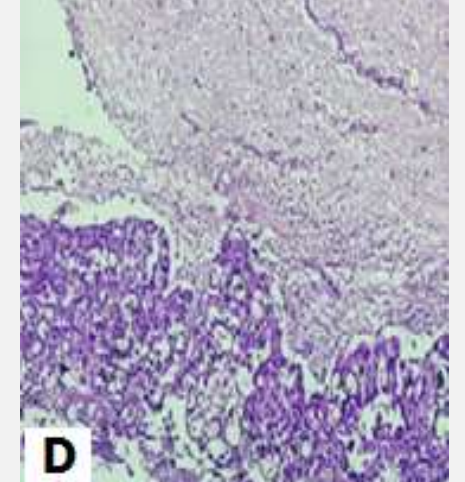
Cerebral metastases are the most common brain tumour seen clinically, comprising slightly more than half of the brain tumours. But the presentation of cerebral metastases as lesions eroding the bone and presenting as bony swellings of the scalp is rare. These cases are studied in our present study. Our study consisted of 4 cases of bony scalp swellings arising from the primary malignant lesion located in each of the cases from lung, breast and thyroid. In one case, the primary site of malignancy could not be detected.

Adenocarcinomatous Metastasis from Lung^{9,10,11}

The initial manifestations of lung carcinoma are cough, haemoptysis, shortness of breath etc. whereas in very few

cases, bony swellings are the first manifestations of lung cancer.¹² In the study done by Kagohashi et al. only 2.3 % of the patients had bony swellings as the initial presentation of lung cancer. Post mortem studies have shown up to an incidence as high as 36 per cent of the bony metastases from lung cancers. The presence of bony metastasis, in general, is a poor prognostic indicator.

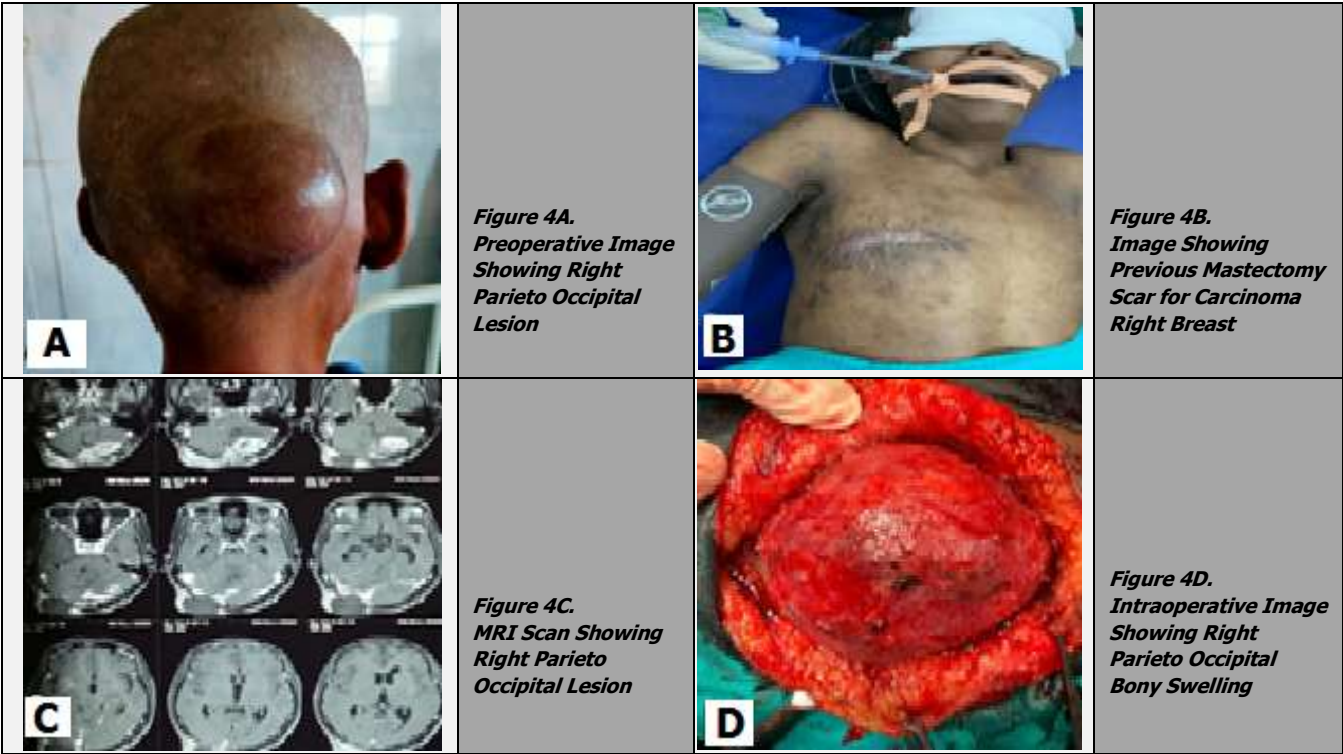
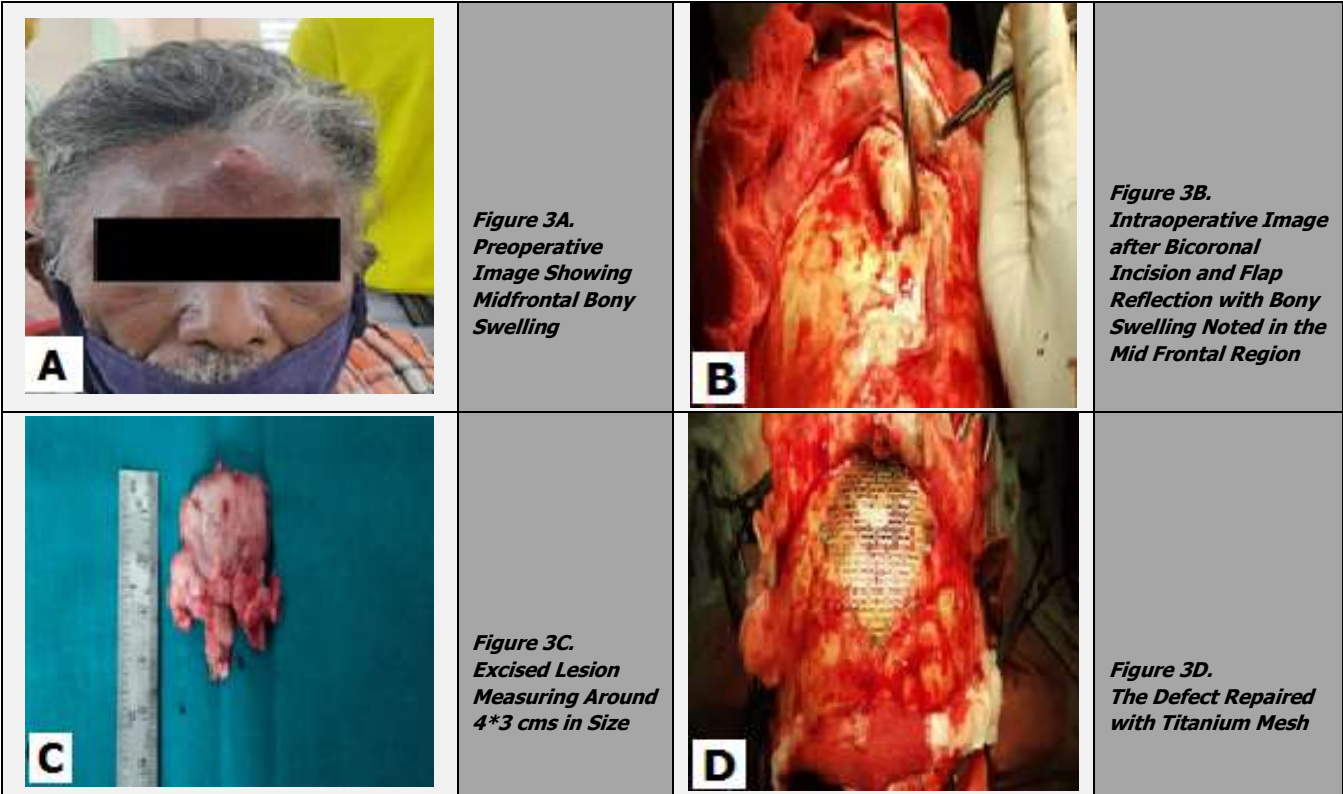
In our study, one patient of age 78 years had presented to us with a bony swelling in the right parieto-occipital area with no other clinical symptoms and not associated with any focal neurological deficits. On preoperative work up on chest x-ray, the patient had a lesion in the left lung. On further investigation, malignancy of the lung was detected. Excision and biopsy of the right parieto occipital lesion showed an adenocarcinomatous metastatic lesion, with the primary being the lung carcinoma.

	<i>Figure 2A. Preoperative Picture of the Patient Showing Right Parieto- Occipital Secondaries with Bony Scalp Swelling</i>		<i>Figure 2B. CT Scan Showing Right Parieto Occipital Region Lesion</i>
	<i>Figure 2C. Excised Right Parieto Occipital Lesion</i>		<i>Figure 2D. HPE Examination Photomicrograph Showing Brain Parenchyma Showing Invasive Tumour Comprising Sheets of Round to Polygonal Cells Exhibiting Moderate Nuclear Atypia. Foci Showing Vague Glandular Formations</i>

Squamous Cell Carcinoma Secondaries from Undetected Primary¹³

Squamous cell carcinoma of the head and neck from an unknown primary site is a well-known entity in literature. The incidence of cancer of undetected primary is stated in the literature between 3 % and 15 %. Imran Kader et al. had presented a case report where the primary squamous cell carcinoma was located in the lung, which produced metastasis in the brain and destruction of parietal bone and with extension into soft tissue mass.¹⁴

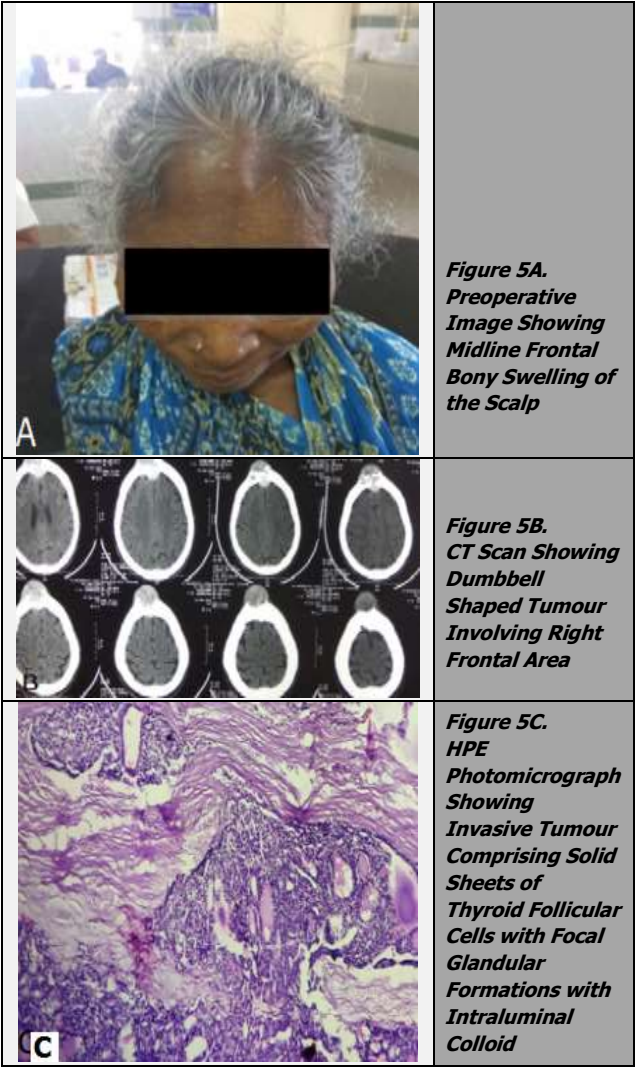
In our study, the patient below is a farmer of age 58 years who had presented with a bony swelling of the forehead in the midline region. There was no complaint of any pain associated with the swelling. He did not have any focal neurological deficits. Preoperative workup was done, and the patient was taken up for surgery. The biopsy of the resected specimen showed metastatic squamous cell carcinoma, but we could not identify the primary site of the squamous cell carcinoma.



Secondaries from Breast Carcinoma¹⁵

Bone is one of the common sites of metastasis for solid tumours like breast malignancy. Our patient is a 69 years old female who had presented with a bony swelling of the right occipito-parietal area. She had no other focal neurological deficits; she had a significant past history of right breast malignancy for which she had undergone modified radical mastectomy of right breast and post mastectomy radiation therapy two years ago. A puckered scar of right modified radical mastectomy is noted, as seen in the images below.

She was thoroughly investigated preoperatively and taken up for surgery. The resected specimen was consistent histopathologically with a metastatic lesion.

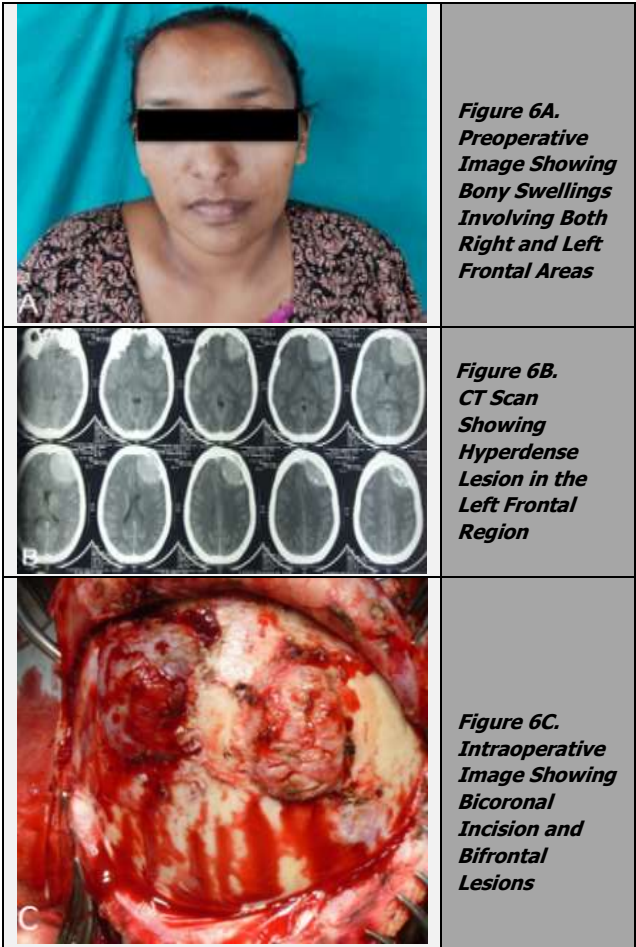


Follicular Carcinoma of Thyroid¹⁶

Follicular thyroid carcinoma (FTC) is a well-differentiated subtype of thyroid carcinoma that has a high propensity for hematogenous metastasis. Follicular thyroid cancers are usually slow growing malignancies and are associated with a good prognosis except when they are presenting with distant metastatic lesions. Skull metastasis from follicular thyroid cancers has well been documented in the literature. Nagamine et al. had reported an incidence of 2.5 % of skull metastasis in thyroid malignancies.¹⁷ Lung and bone are the two most common sites of metastases. In most of the reported cases of FTC metastasizing to the skull, metastases occurred a long time after the diagnosis and establishment of adequate treatment for the primary cancer. Very few cases have been reported with FTC presenting as skull metastasis.¹⁸

The patient below is a 68 years old female who presented with a painless bony swelling on the forehead. The patient had no other clinical symptoms like seizures or focal neurological deficits. She does not have any symptoms suggestive of hypo or hyperthyroidism or symptoms of malignancy like loss of weight or appetite, there was no past history of any radiation. Preoperative workup was done, and excision of the lesion was done. Biopsy showed follicular thyroid pattern on histopathological examination, and on

further investigation, ultrasonography (USG) of thyroid revealed a 1cm mass in the right half of thyroid gland. She was referred to the Department of General Surgery, where she was taken up for surgery, and a total thyroidectomy was done.



Malignant Small Blue Round Cell Tumor^{19,20}

Desmoplastic small round cell tumours are malignant neoplasms of mesenchymal origin. Their most common location is intra-abdominal though they are less frequently seen in other locations like kidneys, lungs, parotid, pancreas, bone or the soft tissues of the extremities and intracranial region also.

In this present case, based upon the clinical and radiological findings, the lesion was strongly suspected to be meningioma but, upon histopathological examination of the excised specimen, the lesion was diagnosed as a malignant small blue round cell tumour. This patient here is a middle-aged female who presented with bifrontal bony swellings of the last one-month duration and had no other complaints like seizures or any focal neurological deficit suggestive of any intracranial pathology. Preoperative workup was done, and she underwent surgery. The histopathological examination of the resected specimen was identified as a malignant small blue round cell tumour.

Malignant small round cell tumours prototypically are composed of cells with round to oval nuclei, scant cytoplasm, and often extensive cellular dissociation. This tumour is histologically and cytologically identical to small cell

carcinoma²¹ originating in other sites with a desmoplastic stroma.

Fibrous Dysplasia

Fibrous dysplasia²² is not a very common entity. It is a non-neoplastic benign developmental disorder of the bone. Fibrous dysplasia is commonly seen in adolescents and young adults in which normal bone is replaced by fibrous connective tissue and immature haphazardly arranged trabeculae of woven bone which is weaker than normal bone and tends to expand. It does not have any gender predilection. It may be monostotic or polyostotic, or syndromic in nature. The monostotic form is the most common and mildest form and accounts for 70 - 80 per cent of the cases. The polyostotic fibrous dysplasia is rare and may be associated with syndromes like McCune-Albright and Mazabraud syndromes. Few consider craniofacial fibrous dysplasia with the involvement of skull and facial bones alone and cherubism involving mandible and maxilla alone as the other subtypes of fibrous dysplasia.

It is an often-incident finding and is usually painless, but it may be revealed by an enlarging mass with symptoms

resulting from mass effect. Calvarial fibrous dysplasia usually crosses the bony sutures, and monostotic lesions may be involving multiple calvarial bones. It preferentially affects the frontal and temporal bones. Bone involvement is focal or multifocal when extensive it typically has a hemispherical preference. Fibrous dysplasia of the bones of the posterior fossa is a rare clinical entity that has been implicated in the process responsible for hindbrain herniation and further leading to the development of syringobulbia.²² Malignant transformation of fibrous dysplasia is rare.

In the present case, a 64 years old male presented with a bony scalp swelling associated with a mild aching type of pain over the swelling, the patient had no other complaints that are suggestive of an intracranial space-occupying lesion, there were no co-morbid conditions in the patient, preoperative investigations were done, and the patient was taken up for surgery, the excised specimen had an intracranial extradural extension of the bony lesion similar to a dumbbell tumour. After surgery, the patient had an uneventful recovery. The histopathological examination revealed abnormal fibrous tissue intermixed with trabeculae of woven bone suggestive of fibrous dysplasia.

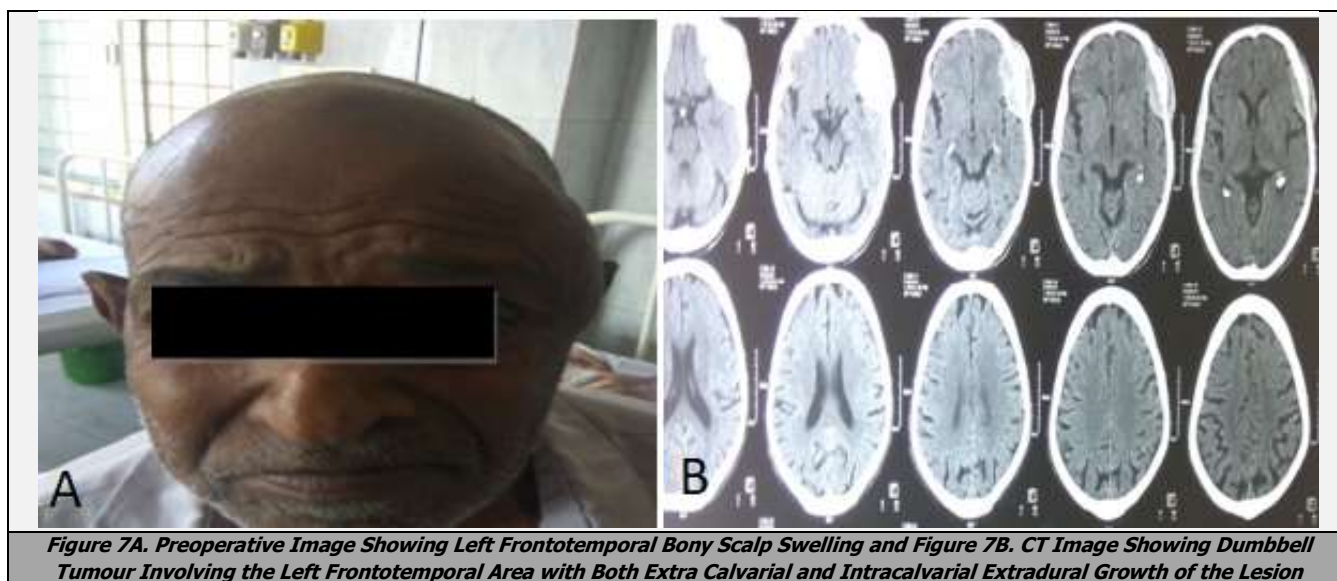


Figure 7A. Preoperative Image Showing Left Frontotemporal Bony Scalp Swelling and Figure 7B. CT Image Showing Dumbbell Tumour Involving the Left Frontotemporal Area with Both Extra Calvarial and Intracalvarial Extradural Growth of the Lesion

CONCLUSIONS

Intracranial lesions eroding the skull and presenting as bony swellings of the scalp are rare. At our institution, in our present study, a total of 13 cases were recorded within a span of two years. All these cases were studied and found to have varied pathological conditions whose spectrum ranges from benign conditions like meningiomas, fibrous dysplasia to malignant lesions like secondaries in the skull from other primaries and malignant small blue round cell tumours.

The management of these lesions has to be done after thorough clinical, radiological, and pathological correlation. The role of pathological characterization of these lesions is of utmost importance in further management that involves radiation therapy and chemotherapy.

Data sharing statement provided by the authors is available with the full text of this article at jebmh.com.

Financial or other competing interests: None.

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