Case Report:
Insular Carcinoma of Thyroid Presenting as a Giant Skull Lesion: A Dilemma in Treatment.

Authors
Rijuneeta Gupta, Associate Professor, Department of Otolaryngology, Head & Neck Surgery,
Ashok K Gupta, Professor & Head (Unit-II), Department of Otolaryngology, Head & Neck Surgery,
Amit Shankar, Senior Resident, Department of Otolaryngology, Head & Neck Surgery,
Sanjdeep Bansal, Assistant Professor, Department of Otolaryngology, Head & Neck Surgery,
Bhattacharya A, Additional Professor, Department of Nuclear Medicine,
Amanjit Bal, Associate Professor, Department of Histopathology,
Post Graduate Institute of Medical Education and Research, Chandigarh.

Address for Correspondence
Dr. Rijuneeta Gupta,
Associate Professor,
Department of Otolaryngology and Head & Neck Surgery,
Post Graduate Institute Of Medical Education & Research,
Chandigarh, India.
E-mail: rijuneeta@yahoo.com

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Abstract: Thyroid surgeons are becoming increasingly more aware of a histologically distinct subset of thyroid carcinoma whose classification falls between well-differentiated and anaplastic carcinomas with respect to both cell differentiation and clinical behavior. This subtype of tumors has been categorized as poorly differentiated or insular carcinoma, based on its characteristic cell groupings. Although the differentiation of insular carcinoma from other thyroid carcinomas has important prognostic and therapeutic significance, relatively little about insular carcinoma has been published in the otolaryngology literature. In this article, we discuss a case of insular carcinoma of thyroid presenting with concurrent distant metastasis to skull, lung, ribs, and inguinal region with review of the literature. We conclude that insular thyroid carcinoma warrants aggressive management with total thyroidectomy and excision of accessible giant lesion followed by radioactive iodine ablation of any remaining thyroid tissue.

Key Words: Insular carcinoma; Thyroid; Metastasis; Treatment.

Introduction
Insular carcinoma first described in 1907 by Langhans as Wuchernde Struma. In 1984, Carcangiu et al described a thyroid neoplasm whose characteristics place it between well-differentiated thyroid carcinoma and anaplastic thyroid carcinoma with respect to its morphologic, biologic, and clinical behavior. Based on its distinct microscopic features this malignancy was labeled as insular carcinoma. Because of insular carcinoma's aggressive nature, proper identification has important prognostic and therapeutic significance. Hence, we present a case of insular carcinoma of the thyroid presenting as a giant lesion over the skull.

Case Report:
A 65 years old woman presented to ENT services of postgraduate institute of medical education and research, Chandigarh with swelling over the occipital region for 1 yr which was progressively increasing in size since two months only. There was no associated hoarseness, dysphagia, respiratory distress or radiation exposure. On examination, the swelling was firm, nontender, immobile, measuring 10x9 cm over the occipital region. Fine needle aspiration cytology from the swelling revealed metastatic well differentiated follicular carcinoma and subsequently from thyroid gland revealed follicular carcinoma. Thyroid function tests were normal. X-ray skull was suggestive of erosion of inner and outer table of occipital bone with overlying soft tissue swelling. MRI brain suggested intracranial extradural extension of tumour. Whole body I-131 scan showed widespread functioning metastasis in neck, skull, chest, ribs and left posterior acetabular margins. Sestamibi tumor imaging after i.v. 99m Tc sestamibi showed increased uptake in occiput, left chest, left inguinal region and in total likely to be metastasis. USG abdomen could not rule out liver metastasis due to fatty liver. Total thyroidectomy was carried out followed by excision of 10x9 cm scalp lesion. However, histopathology from thyroid specimen was poorly differentiated carcinoma which was in correlation with histopathology of scalp lesion i.e solid sheets of...
microfollicles. Both reports were reviewed and it revealed insular carcinoma poorly differentiated. Post surgery, sestamibi scan showed foci of tracer uptake at skull, ribs, inguinal region and lung. After three months of follow up, patient was then subjected to radioiodine (120 mci $^{131}$I) for ablation of residual lesion. Now the patient is on regular follow up for the last 2 years having well healed scalp wound and is taking thyroid supplement therapy.

Fig 1: NCCT Head showing a large skull metastatic lesion causing bone destruction

Fig 2: MRI Brain showing extradural occipital metastasis

Fig 3: X-Ray Chest showing rib metastasis

Fig 4(a) & (b): $^{131}$I scan showing uptake in chest rib and pelvis
Discussion

Insular carcinoma of the thyroid is an uncommon pathologic entity, the reported incidence of insular carcinoma in the literature ranges widely, from 0.4 to 10% of all thyroid cancers. Insular carcinoma is a relatively new distinct subtype of thyroid carcinoma and the criteria for diagnosis is not uniform among pathologists. Well-differentiated carcinoma can progress to insular carcinoma and then to anaplastic carcinoma of the thyroid by dedifferentiation. Distant metastases in insular carcinoma of the thyroid were often noted in the lungs and bones. In our patient, metastases were seen in the skull and inguinal region. The microscopic features as described by Carcangiu et al include solid clusters “nests” of tumor cells containing a variable number of follicles, often sharply separated by artifactually created clefts. Other critical features are small size and uniformity of tumor cells, variable but consistent mitotic activity, necrosis with capsular and vascular invasion which at times lead to the formation of peritheliomatous structures. The diagnosis of insular carcinoma is often elusive because these tumors have been classified as poorly differentiated papillary carcinomas when papillary features are present and as undifferentiated thyroid carcinomas of the small-cell category when papillary features are absent.

Diagnosis of insular carcinoma by fine needle aspiration cytology has been reported but it can be complicated if a cell sample is obtained from a tumor component other than insular carcinoma. Only definitive way to diagnose thyroid insular carcinoma is by histologic examination of the excised tumor.

Insular carcinoma has an intermediate position between well-differentiated carcinoma and anaplastic carcinoma with regard to both histologic features and biologic aggressiveness. It has been shown that the extent of poorly differentiated components in a well differentiated thyroid tumor can affect the prognosis, tumors with >10% of poorly differentiated components are associated with frequent regional recurrences, distant metastasis and poor prognosis. Some authors found that insular carcinoma was associated with a worse prognosis than that of well-differentiated carcinoma. Insular carcinoma of the thyroid has better prognosis, and aggressive treatment is beneficial to the patients. Ashfaq et al concluded that insular carcinoma within papillary or follicular carcinoma did not adversely affect the patient's prognosis and that the only significant factors that affect the behavior of insular thyroid carcinomas were advanced patient age and the tumor stage at diagnosis. Most common treatment of insular thyroid carcinoma was total thyroidectomy with ablation of remaining functioning thyroid tissue by radioactive iodine. With over 50% of poorly differentiated thyroid carcinomas having regional nodal metastases, with thyroidectomy and possible modified...
radical neck dissection should be considered. The current protocol of management is total thyroidectomy along with excision of accessible distant metastatic lesion followed by adjuvant treatment (radioactive iodide) and close follow-up. Justin et al recommended postoperative imaging for the early detection of persistent disease or metastasis in order to enhance survival or palliation. However, in their retrospective review of 25 cases, Carcangiu et al reported that neither the extent of the surgical operation nor the prophylactic use of radiiodine altered the course of disease or the rate of cervical or distant metastasis. External-beam radiation was used in patients with invasive tumors, persistent local disease, or bony metastases. Recurrences were often local, but lung and bony metastases were also common. Typically, the tumor's insular pattern is conserved in recurrences or metastases.

Conclusion
Insular carcinoma is characterized by a distinctive histologic appearance and an aggressive clinical behaviour. The propensity for local recurrence and distant metastasis mandates aggressive therapy at the time of initial diagnosis. It is important for clinicians to be able to recognize and differentiate this entity for appropriate management of this tumor.

References