ISSN (O):2395-2822; ISSN (P):2395-2814

Tender Fast Growing Swelling of the Left Maxilla.

Asha Karadwal[1], Swati Roy[2], Sonam Bhalla[3], Mohit Kapoor[1], Rohit Sharma[2], Priyanka Sharma[4]

[1]lecturer, Department of Oral Pathology, Yamuna College of Dental Sciences and Research, Yamunanagar (Haryana), India.
[2]Reader, Department of Oral Pathology, Yamuna College of Dental Sciences and Research, Yamunanagar (Haryana), India.
[3]Lecturer, Department of Oral Pathology, Yamuna College of Dental Sciences and Research, Yamunanagar (Haryana), India
[4]lecturer, Department of paediatric dentistry, Yamuna College of Dental Sciences and Research, Yamunanagar (Haryana), India.

Received: September 2017
Accepted: September 2017

ABSTRACT

A 63 year old male reported with mild pain and gradually enlarging swelling of presumed dental origin in the left side of the face since two months. Based on clinical and radiographic finding, the differential diagnosis for this lesion included squamous cell carcinoma, nasopharyngeal carcinoma, salivary gland malignancies, sinonasal carcinoma, non hodgkin lymphoma large B cell type and deep fungal infection. Biopsy was taken and microscopic examination revealed features of sinonasal carcinoma.

Keywords: Carcinoma, malignancy, maxilla.

INTRODUCTION

A 63 year old male reported with mild pain and gradually enlarging swelling of presumed dental origin in the left side of the face since two months. The patient's medical history revealed that he had taken antibiotics and analgesics for pain and swelling but had no relief. Extraorally, on inspection, the swelling extended superioinferiorly from infraorbital margin to the angle of lip and anterioposteriorly from ala of the nose to the zygomatic prominence with ill defined margins. Consistency of swelling was firm and there was slight tenderness on palpation. Intraorally, soft tissue obliteration of vestibule in respect to maxillary molar region was observed. Paranasal radiograph revealed destruction of left maxilla and zygoma with a soft tissue opacity filling the complete left maxillary sinus [Figure 1]. Computed tomography revealed heterogenous enhancing mass lesion involving left maxillary sinus and a breach in the anterior and posterior wall of the sinus with extension of soft tissue anteriorly and laterally into the infratemporal fossa [Figure 2].

Differential Diagnosis

Based on clinical and radiographic finding, the differential diagnosis for this lesion included squamous cell carcinoma, nasopharyngeal carcinoma, salivary gland malignancies, sinonasal carcinoma, non hodgkin lymphoma large B cell type and deep fungal infection.

Malignancy arising within the paranasal sinuses is relatively rare, constituting 1.0% of all malignancies, with approximately 80% of these malignancies arising in the maxillary sinus with a lesser prevalence in the ethmoid sinus. Malignant disease of the sphenoid and frontal sinuses is very rare. Almost 80% of malignancies are squamous cell carcinomas, with acinic cell carcinomas forming 10% of the sinus malignancies.[1] The Squamous cell carcinoma of the nasal and paranasal region has been reported to occur most frequently in the maxillary sinus (55-60%) followed by nasal cavity (19-35%), ethmoid sinus (9-15%), nasal vestibule (4%) and frontal and sphenoid sinus (1% each).[2] Therefore, it may present with any one of many signs and symptoms including nasal stiffness, swelling over the cheek, pain in the area and epistaxis. It usually presents as a mass bulging into the oral cavity or into the nasal cavity. Because it erodes the sinus floor, early mobility of the maxillary teeth may be a presenting sign. Radiographically, it reveals a soft tissue mass in the maxillary sinus and usually bony erosion of one or more walls of the sinus. Microscopically, poorly differentiated squamous cell carcinomas usually show small areas of obvious squamous differentiation or keratinisation.[3]

The majority of nasopharyngeal carcinoma arise from the lateral nasopharyngeal wall (fossa of Rosenmuller) and frequently extend to the paranasopharyngeal space, from which the tumor

Name & Address of Corresponding Author
Dr. Asha Karadwal,
M.D.S.
House no 1185 Suthran street town hall
jagadhri 135003,
Haryana, India.

may grow along the preneurial space of the trigeminal nerve. It accounts for 75% of malignancies in the nasopharynx and represents a rapidly proliferating and metastasizing type of cancer of squamous cell origin. The age incidence curve is bimodal, with a peek occuring between 15 and 25 years and another between 60 and 69 years. Nasopharyngeal carcinoma has a strong male predilection with a men being three times more commonly affected then women. Most patients with signs or symptoms of a tumor mass with nasal obstruction, epistaxis, dysphagia and pharyngeal pain. The first sign of the disease for 50-60% of patients is an enlarged, firm to hard, cervical lymphnode, which represents metastatic tumor. The tumor may invade through the foramen lacerum into the brain, producing central nervous system symptoms or it may involve the cranial nerves. 5% - 10% of patients also have distant metastasis at the time of diagnosis. Most nasopharyngeal carcinomas are squamous cell carcinomas that arise from respiratory mucosa. Undifferentiated carcinoma (also known as lymphoepithelioma- like carcinoma) is a clinically and morphologic distinctive variant of squamous cell carcinoma. Microscopically, It composed of cells with indistinct borders, vesicular nuclei with inconspicuous nucleoli and often show a marked lymphoplasmacytic infiltrate (lymphoepithelioma). The tumor cells in undifferentiated carcinoma demonstrate a syncytial growth. Cytological atypia is less prominent and necrosis is not usually seen.[3,4]

Salivary gland malignancies reportedly constitute 4-8% of neoplasms of the sinonasal tract. These tumors arise from the serous mucinous glands of the nasal cavity and paranasal sinuses and the overlying surface epithelium. The largest of the paranasal sinuses are the maxillary sinuses. These triangular cavities are located in the body of each maxillae. Of the paranasal tumors, the majority are found in the antrum. Most tumor of the paranasal sinuses are malignant, adenoid cystic carcinoma is the most common variety. Several other types of salivary gland tumors have been reported in the sinonasal region including mucoepidermoid carcinoma, acinic cell carcinoma.[5] In contrast Bhattacharya in his series of 188 cases had adenoid cystic carcinoma (34%) as the predominant histological type followed by sarcomas (24%).[6] Sinonasal malignancies occur twice as often in males as in females, and are most often diagnosed in patients 50 to 70 years of age. Mucoepidermoid carcinomas arising from mucous glands of maxillary sinus are extremely rare and account for 13% of all malignancies occurring in maxillary sinus.[7] Microscopically, salivary gland malignancies are characterized by a biphasic pattern of squamous nests and duct like structure at the periphery of the lesion that blend into solid nodules centrally.[5]

Sinonasal carcinoma is an unusual tumor responsible for less than 1% of cancer deaths as reported by United States.[5] It is a highly aggressive, undifferentiated anaplastic carcinoma, without obvious squamous or glandular differentiation. The age range is broad and includes young adults and elderly people with the median age at diagnosis being the sixth decade.[8] A marked left sided preponderence has been noted for ethmoid tumors, suggestive of an exogenous pathogenesis. Sinonasal carcinoma often are diagnosed late in their course, when extensive bone destruction is already present.[5] It presents as a rapidly enlarging tumor mass involving multiple (sinonasal tract) sites, often with evidence of extension beyond the anatomic confines of the sinonasal tract. It most commonly involves the nasal cavity, maxillar sinus, ethmoid sinuses, orbit and cranial cavity. The tumor affects males more often then the females and has a broad age range with a male to female ratio of approximately 2:1 to 3:1. Symptoms including epistaxis, proptosis, visual anomalies, cranial nerve deficits, swelling and pain. Cranial nerve palsies are a common finding as well. Symptoms are related to an extensively infilterative and destructive sinonasal mass. Radiographic examination typically reveals a large, expansile sinonasal mass with bony destruction and invasion of adjacent structures.4 Microscopically, sinonasal undifferentiated carcinoma is classically composed of small to medium-sized undifferentiated cells and is characterized by high mitotic rates, significant cellular pleomorphism, and high nuclear to cytoplasmic ratios, necrosis and vascular invasion.[9]

Non-Hodgkin's Lymphoma are a group of neoplasms that originate from the cells of the lymphoreticular system. 40% of Non-Hodgkin's lymphoma arises from extranodal sites. The nasal cavities and paranasal sinuses are rarely affected by primary non-Hodgkin's Lymphoma. Common primary extranodal sites of lymphomas include stomach, liver, soft tissue, dura, bone, intestine and bone marrow. Most patients present with rapidly enlarging masses, often with symptoms both locally and systemically (fever, recurrent night sweats, or weight loss). According to Danish study in 1977 showed that non-Hodgkin lymphomas were formed the second most common malignant tumors in the sinonasal region, with squamous cell carcinoma being the most common. The majority of sinonasal lymphomas are the B-cell type, the most common histologic type being diffuse large B cell lymphoma. Large B cell lymphoma is a fast growing malignancy that may arise inside or outside of the lymphatic system. A wide histologic and biological spectrum of B cell lymphomas occur

in the head and neck. In large B cell lymphoma, there is a slight male predeliction. Most tumors are locally advanced at presentation, with frequent bone destruction and extension into the adjacent sinuses, nasopharynx or palate. Within bone, the tumor produces extensive destruction. Microscopically, non hodgkin lymphoma large B cell type is composed of sheets of large lymphoid cells showing abundant cytoplasm and nuclei comparable in size or larger than reactive histiocytes. Within lymphnodes, normal lymphoid architecture is effaced and necrosis is common.[10]

Although fungal infections of the paranasal sinus are uncommon, 3–5% of incidence is reported. Mucor and aspergillus species are the most common causative agents of fungal sinusitis, but infection with lesser known species have been reported across the world infrequently.[11] Inflammatory sinus disease such as chronic sinusitis with granulation tissue accumulation may also produce the same signs and symptoms such as nasal stuffiness and a mass related enlargement of the maxilla. Mycotic infection of the upper respiratory tract are usually located in the paranasal sinuses. Mucormycosis is one of the most frequent. Mucormycosis is an opportunistic infection affects immunocompromised patients with uncontrolled diabetes, leukemia and renal failure. It is caused by organisms of one of the three genuses of the family mucorales: absidia, mucor and rhizopus. Pain and swelling precede ulceration, exposing grey black necrotic nasal cartilage, vomer and ethmoid bones. Tissue necrosis may result in perforation of the palate. Extension into the orbit or brain is a common complication. The fungus has a propensity for arterial wall invasion, which may lead to hematogenous spread, thrombosis, or infarction. Radiographically sinus and orbital extensions are recognized by membrane or periosteal thickenings as well as bony disruption. Another microorganism responsible is aspergillus. The first presentation of aspergillus is an external otitis and second presentation is usually a chronic maxillary sinusitis, particularly when previous surgeries have disturbed the normal physiology and blood supply of the sinus and even more commonly when the foreign bodies have been placed in the sinus and patient complaints nose stuffiness in the nose and sinus, chronic pain and intermittent nasal drainage. In severely immunocompromised, aspergillosis may present as a necrotic black skin ulceration on the face or neck or as sinus infections, resembling mucormycosis.[12] Foci of infection may lead to dystrophic calcification and the formation of rhinoliths, which may be seen on dental radiographs. Large rhinoliths are known as fungal balls.[1] Radiological evidence of invasive infection in the sinuses (i.e., erosion of sinus walls or extension of infection to neighbouring structures, extensive skull base destruction) may present. Microsopically, mucormycosis, an acute and chronic inflammatory infiltrate is seen in response to the fungus. The organism is usually readily identified in hematoxylin and eosin stained sections in areas of tissue necrosis. Characteristic necrotic vessel walls containing thrombi and fungi may be evident. Microscopically, the fungus consists of large, pale-staining, non-septate hyphae that tend to branch at right angles. Mucormycosis is characterized by the presence of wide, flat non-septate fungal organisms. Aspergillus appears as branching septate hyphae, invades blood vessels, causing occlusion and consequent necrosis.[12]

DIAGNOSIS

A biopsy of the involved area was necessary to rule out the possibility of an underlying malignant process. An incisional biopsy under local anesthesia with respect to posterior left buccal vestibule of maxilla was performed. On incisional biopsy, soft tissue was removed in abundant depth till the bone and perforation (bony extension) of tissue was observed and tissue was found to be proliferating outward from the maxillary sinus. Soft tissue specimens from left buccal vestibule of maxilla and hard tissue specimens from left posterior maxilla were taken for histopathological examination.

Microscopic examination of the specimen revealed presence of nests, trabeculae and sheets of medium sized malignant epithelial cells with high mitotic rate. In some areas the peripheral layer of tumor cells resemble cylindrical cells. The tumor cells are large, highly anaplastic with vesicular nuclei and single or multiple nucleoli. Extensive necrosis is evident throughout the tumor mass. These features are suggestive of sinonasal carcinoma because microscopically no evidence of lymphoplasmacytic infiltrate, squamous differentiation or keratinization, sheets of large lymphoid cells and fungal hyphae were seen [Figure 3 & Figure 4]

TREATMENT

The patient is undergoing combined chemotherapy and surgical treatment. Chemotherapy prescribed injection Paclitaxel 250mg and carboplatin 450mg, rago D capsule (rabeprazole sodium 20mg + domeperidone 30mg), tablet niap (niap paracetamol 500mg + nimesulide 100mg), dynapar (paracetamol 500 mg), tryptomer (amitriptyline 10mg) and otrivin (xylometazoline) 0.1% w/v.

DISCUSSION

Sinonasal undifferentiated carcinoma is a rare, highly aggressive and clinico-pathologically distinctive carcinoma of uncertain histogenesis.

The disease affects males more often than females and has a broad age range. Sinonasal undifferentiated carcinoma presents as a rapidly enlarging tumour arising from the sinonasal tract with initially vague symptoms that are of relatively short duration. It most commonly involve the nasal cavity, the maxillary antrum and the ethmoid sinuses. In the vast majority of cases, patients present with pain and swelling of the jaws with tumour expansion through a defect in the anterior–facial wall of the left maxillary sinus. In addition, it frequently invades into the surrounding structures, including the nasopharynx, orbit and cranial cavity. Symptoms are due to the large size and local invasion and include epistaxis, proptosis, visual anomalies, cranial nerve deficits, and pain. Symptoms are of relatively short duration.[13]

Pathological examination of Sinonasal undifferentiated carcinoma typically reveals large tumours with fungating and poorly defined margins that invade adjacent structures. The histologic appearance is characterized by sheets, trabecular and ribbon-like arrangements of small to medium-size undifferentiated cells. These cells often have high nuclear to cytoplasmic ratio, high mitotic rate and prominent tumour necrosis. Lymphovascular and neural invasion are often also identified.[8]

It is a malignant tumour of the sinus (sinonasal) that is of epithelial origin (carcinoma), but lacks evidence of keratin production (undifferentiated). Immuno-histochemical antigenic profiles vary widely. The majority of patients present with advanced stage disease and often undergo intense, multi-modality treatment. Unfortunately, survival remains poor. Due to the small number of cases seen, the ideal treatment regimen has not been systematically evaluated. However, treatment generally involves surgical removal of the tumour. Patients with sinonasal undifferentiated carcinoma have a high rate of both local–regional recurrence and distant metastasis. Moreover, because of the complex anatomy of the head and neck area, complete removal of the tumour with wide margins is not always possible. Consequently, surgery is commonly combined with radiation or chemotherapy or both.[14,15]

CONCLUSION

Unfortunately, the prognosis for a patient with sinonasal undifferentiated carcinoma is extremely poor. Most patients die of local disease within 1 year of diagnosis. Nevertheless, long-term survival has been documented in some patients, especially in cases diagnosed early in the disease process.

REFERENCES

1. Bell GW, Joshi BB, Macleod RI. Maxillary sinus disease: diagnosis and treatment. Br Dent J 2011;210(3):113-8.
2. Slootweg PJ, Richardson M. Squamous cell carcinoma of the upper aerodigestive system. In: Gnepp DR, editor. Diagnostic surgical pathology of the head and neck. 2nd ed. Philadelphia: W.B. Saunders Co. (Elsevier); 2009. p. 54-60.
3. Marx RE, Stern D. Premalignant and malignant epithelial tumors of mucosa and skin. In: Hauffman L, editor. Oral and maxillofacial pathology A rationale for diagnosis and treatment, vol 1. 2nd ed. Chandler Drive Hanover Park: Quintessence pub co, Inc; 2012. p. 351-354.
4. Neville BM, Damm DD, Allen CM, Bouquot JE. Epithelial pathology. In: Neville BM, editor. Oral and maxillofacial pathology 3rd ed, W.B. Saunders Co. (Elsevier) St. Louis, Missouri. 2009. p. 362-452.
5. Rosai J, editor. Rosai and Ackerman's Surgical pathology, Vol. 1. 9th ed. St. Louis, Missouri: Mosby (Elsevier); 2005. p. 305-334.
6. Bhattacharyya N. Survival and staging characteristics for nonsquamous cell malignancies of the maxillary sinus. Arch otolaryngol Head Neck Surg. 2003;129(3):334-7.
7. Daryani D, Gopakumar R, Nagaraja A. High grade mucoepidermoid carcinoma of maxillary sinus. J Oral Maxillofacial Pathol 2012;16(1):137-40.
8. Wenig BM. Undifferentiated malignant neoplasms of the sinonasal tract. Arch Pathol Lab Med 2009;133(5):699-712.
9. Enepekides DJ. Sinonasal undifferentiated carcinoma: an update. Curr Opin Otolaryngol Head Neck Surg 2005;13(4);222-5.
10. Prasad ML, Perez-Ordonez B. Nonsquamous lesions of the nasal cavity, paranasal sinuses, and nasopharynx. In: Gnepp DR, editor. Diagnostic surgical pathology of the head and neck. 2nd ed. Philadelphia: W.B. Saunders Co. (Elsevier); 2009. p. 172-175.
11. Rai S, Tiwari R, Sandhu SV, Rajkumar Y. Hyalohyphomycosis of maxillary antrum. J Oral Maxillofac Pathol 2012;16(1):149-52.
12. Marx RE, Stern D. Infection diseases of the oral and maxillofacial pathology. In: Hauffman L, editor. Oral and maxillofacial pathology A rationale for diagnosis and treatment, vol 1. 2nd ed. Chandler Drive Hanover Park: Quintessence pub co, Inc; 2012. p. 111-114.
13. XU CC, Dziegielewski PT, McGaw WT, Seikaly H. Sinonasal undifferentiated carcinoma: the Alberta experience and literature review. J Otolaryngol Head Neck Surg 2013;42(1):2.
14. Goel R, Ramalingam K, Ramani P, Chandrasekar T. Sinonasal undifferentiated carcinoma: A rare entity. J Nat Sci Biol Med 2012;3(1):101-4.
15. Edwards PC, Hess SJ, Saini T. Sinonasal undifferentiated carcinoma of the maxillary sinus. J Can Dent Assoc 2006;72(2):163-7.

How to cite this article: Karadwal A, Roy S, Bhalla S, Kapoor M, Sharma R, Sharma P. Tender Fast Growing Swelling of The Left Maxilla. Ann. Int. Med. Den. Res. 2017; 3(6): DE25-DE28.

Source of Support: Nil, **Conflict of Interest:** None declared