A CASE OF EWING SARCOMA OF THE MAXILLA

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CASE REPORT

A 23-year-old female consulted due to a slowly enlarging right maxillary mass that initially presented as recurrent epistaxis for 6 months, occurring 2-3x/week amounting to 1 tablespoon which was resolved by finger pressure. The mass was only noted 1 month prior measuring 1x1 cm and was associated with headache and thick yellowish, non-foul-smelling nasal discharge. On PE, mass was noted to be firm, slightly movable, tender on deep palpation, and measures 4x3cm in the right maxillary area (Fig. 1). Nasal endoscopy revealed a mass occupying the right nasal cavity (Fig. 3). Contrast-enhanced CT scan of the paranasal sinuses showed a lesion of the anterior wall of the right maxillary sinus extending to the medial wall (Fig. 4). Endoscopic biopsy showed a CD99-positive malignant round cell neoplasm (Fig. 5). These support the diagnosis of an extraskeletal Ewing Sarcoma/ PNET. The mass (Fig. 2) was resected with preservation of the orbital floor and hard palate. The biopsy of the mass confirmed Ewing Sarcoma of the maxillary sinus, right with negative margins on all sides.

DISCUSSION

Bone malignancies account for only 0.19% of all reported cancer cases as of 2018 (1). Of these, Ewing sarcoma accounts only to 4-6% of all primary cases (2). Usually found in the bones and soft tissues, only 1-4% occurs in the bones of the facial area (mostly in the mandible and calvaria) and rarely involves the paranasal sinuses (3).

Masses in the maxillary sinus usually remains undetected until it protrudes to the nasal/oral cavity producing signs and symptoms like nasal obstruction, epistaxis, destruction of the palate and ulceration of the overlying mucosa as with our patient. This delay can be attributed to the volume the maxillary sinus can contain which allows expansion of the mass before detection (4).

In diagnosis, CT Scan is utilized in detecting bone changes/damage while MRI is more often used in staging since it can reveal tumor extension to adjacent structures. The radiologic signs include expansion and erosion of the cortical bones, as seen with the destruction of the walls of the maxilla in this case (5). Histologic features of the biopsy includes presence of small round cells, nucleus with scanty cytoplasm and fine chromatin with occasional Homer-Wright rosettes. Its reactivity to CD99 and absence of reaction to other markers (CK, LCA, CD3, CD20) confirmed our diagnosis (6).

In this patient, we widely resected the mass via a Weber-Ferguson Dieffenbach incision before the initiation of chemotherapy because concerns of waiting for the chemotherapy can cause significant delay that may lead to further extension of mass to adjacent structures, thus requiring a more aggressive approach. Chemotherapy commenced shortly after the procedure.

CONCLUSION

Due to its rarity, Ewing Sarcoma of the maxilla requires skillful evaluation of the history, PE, and ancillary procedures in order to recognize, diagnose and treat the disease in its early stages. The management applied to this patient may be an option for earlier stages of the disease in which good margins of resection can be achieved, especially if initiation of chemotherapy will be delayed leading to invasion of more adjacent structures, hence leading to a poorer prognosis. However, more evidence regarding this management is necessary.