Isolated Orbital Metastasis in Occult Rectal Adenocarcinoma - A Case Report

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ABSTRACT

Introduction: Orbital metastasis is a rare disease, though it may be the first manifestation of a primary tumor in up to 30% of cases. In adults, metastasis is often from carcinoma breast, lung and melanoma. **Case report:** A 23 year old male developed swelling around the outer aspect of the right eye. Post operative histopathology stated the possibilities as small cell variant of osteogenic sarcoma or ewings sarcoma/PNET of the right eye. IHC favoured metastatic deposit of poorly differentiated adenocarcinoma. PET CT and Colonoscopy confirmed the diagnosis of carcinoma rectum. Bone scan revealed orbital metastasis.

Conclusion: Carcinoma of the rectum with orbital metastasis is a very rare presentation. However, we must consider this association in order to recognize the onset and administer appropriate treatment. The authors believe that this is a rare case and the first one reported with presentation first as an orbital metastasis with subsequent diagnosis of the occult primary.

Key words: carcinoma rectum, orbital metastasis

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INTRODUCTION

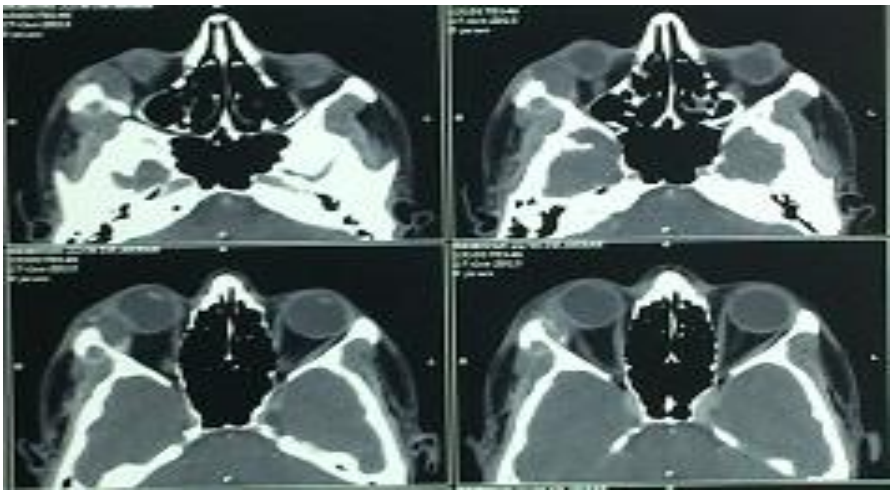
Orbital metastasis is a rare disease, accounting for less than five percent of all orbital conditions. Although the diagnosis of the primary tumor usually precedes the diagnosis of orbital metastasis, it may be the first manifestation of a primary tumor in ups to 30% of cases, hence the interest of early recognition. In adults, metastasis is often from carcinoma breast, lung and melanoma. Gastrointestinal tumors, renal tumors, thyroid tumors, pancreatic cancer and prostate cancer are rare.

Orbital metastasis from colorectal cancers is extremely rare. Proptosis is the most frequent presenting clinical sign. Once the diagnosis is made, the prognosis is poor and the treatment is palliative.

CASE REPORT

A 23 year old male presented with a one and a half month history of swelling with pain around the outer aspect of right eye. Patient had no other complaints. On examination, there was a hard, fixed swelling of lemon size located around the lateral canthus of the right eye. Systemic examination was normal. Ophthalmic examination showed unilateral ptosis, oedema and abductive restriction. CT scan was suggestive of soft tissue mass centered on right orbital wall with osteolysis extending into right orbit. There was no intracranial extension or extension into the infra temporal fossa. FNAC was suggestive of PNET/ Ewing's sarcoma.

Figure 1: CT scan axial slices showing right orbital metastasis



After excision of right orbital mass, histopathological examination stated the possibilities as Small cell variant of osteogenic sarcoma and Ewing's sarcoma/PNET of the right eye. On performing IHC, CD99 and Vimentin were negative, ruling out the possibility of round cell osteogenic sarcoma/ Ewing's sarcoma. Pancytokeratin was diffusely positive and CD45 (LCA) was negative in the tumor cells. Lateral orbital bone histomorphology and pancytoketarin IHC positivity was in favour of metastatic deposit of poorly differentiated adenocarcinoma.

Post surgery, patient had complaints of progressively increasing painful swelling at the same site. On performing a whole body PET CT Scan, CT findings revealed a destructive lesion associated with a soft tissue component involving lateral wall of right orbit, zygomatic arch and body of zygomatic bone. The proximal rectum showed a circumferential enhancing mass narrowing its lumen, extending from the recto-sigmoid junction to mid rectum for a length of 3.5cm. The mass extended into perirectal fat as stranding. PET findings revealed markedly increased uptake in the right orbital wall and zygomatic lesion and the rectal lesion as described on CT. There was increased uptake in left supraclavicular and abdominal nodes. The final impression on PET-CT was suggestive of rectal tumor with nodal and skeletal metastasis.

Figure 2: Patient presenting with diffuse swelling in the right periorbital area

Colonoscopy revealed a stenotic growth in the rectum 6cm from the anal verge. Colonoscopic biopsy was reported as poorly differentiated adenocarcinoma of the rectum. On performing a Tc99 bone scan, abnormally increased tracer uptake was noted in right maxilla and lateral orbital wall, consistent with metastasis. The final diagnosis of carcinoma rectum with orbital metastasis was made and patient was started on palliative chemoradiotherapy.

DISCUSSION

The orbit is a rare site of distant metastasis with incidence varying from 1% to 13% in the reported series of all orbital tumors [1]. The diagnosis of orbital metastasis is often unexpected when an ophthalmic manifestation is the first presentation. This is more frequently seen with lung, gastrointestinal, thyroid and renal carcinomas [1,2] . In contrast, the majority of breast cancers with ocular metastasis have had treatment for the primary tumor before the eye becomes involved [3,4] . In upto 35% of the cases, the primary neoplasm remains unknown despite systemic evaluation or autopsy [1,2] . Most metastatic carcinomas of unknown origin are from the lung or pancreas.

Unlike other primary orbital expansive processes, metastases usually have a rapid growth with early onset of symptoms, and in most cases are unilateral. Presenting symptoms are usually diplopia (48%), proptosis (26%) or pain, but we can also find decreased visual acuity (16%), decreased motility, palpable mass, periorbital swelling or ptosis. Approximately 5% of the orbital metastasis involve the extraocular muscles, and they are usually unilateral, unlike choroidal metastasis. Both CT and MRI are essential and more useful than ultrasound for evaluating orbits with suspected metastatic lesions.

The prognosis in patients with orbital metastasis is often poor as the primary tumor can only be found in one third of the patients. The median survival is reported as a little over 1 year, only 27% have a 2-year survival. Management is based on establishing a correct diagnosis, the systemic status of the patient and whether optic nerve compression is present.

Of the six reports of colorectal adenocarcinoma, only 3 contained illustrated orbital histopathology, two of which were well differentiated

adenocarcinoma and signet ring cell carcinoma [5-7].

The other report, claiming a metastasis of mucinous adenocarcinoma to the breast and orbit from colon, has not excluded breast as the primary source of cancer [8].

Since this metastasis usually present in advanced stage, treatment is usually palliative, by I.V steroids and specially radiotherapy, to decrease tumor size and consequently, the patient's pain. In general, patients with orbital metastasis are not candidates for orbital surgery for the removal of tumor mass, since this does not lead to cure of the disease. However, in some cases in which the tumor growth is slow, the removal of metastasis with the primary tumor may improve prognosis.

CONCLUSION

Carcinoma of the rectum with orbital metastasis is a very rare presentation. However, we must consider this association in order to recognize the onset of clinical orbital metastasis and therefore administer appropriate and early palliative treatment to improve the quality of life in these patients.

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