Isolated orbital metastasis from Adrenal cortical carcinoma – A rare case report with Review of literature

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ABSTRACT

Introduction: Adrenal cortical carcinoma (ACC) is a rare tumor which has an aggressive disease course and an early metastasis. When present, metastasis mainly involves lung or bones. Orbital metastasis from adrenal cortical carcinoma is a rare occurrence. The present report describes a case of adrenal cortical carcinoma with isolated orbital metastasis. **Case presentation:** A 54-year-old postmenopausal women who was operated for adrenal cortical carcinoma 1 year back presented with protrusion of left eye which was diagnosed as isolated metastasis to orbit after complete work up. **Conclusion:** ACC is a rare and an aggressive tumour with poor long term survival which infrequently spreads to the orbit. The current treatment protocols are disappointing and there is a need for better therapies. Further studies are needed at molecular level to understand the pathogenesis of the disease and to give better outcome.

Keywords: Adrenal cortical carcinoma, orbital metastasis, Post menopausal women

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INTRODUCTION: Adrenal cortical carcinoma (ACC) is a rare and heterogeneous malignancy with incompletely understood pathogenesis and poor prognosis. The tumor is aggressive and metastasizes early in the disease course. Patients often have metastatic disease at initial presentation, with the most common sites being liver, lung, lymph nodes, and bone. [1] Spread to orbit is rare. We here in report a case of ACC with isolated orbital metastasis.

CASE REPORT: A 54-year-old postmenopausal women presented with history of persistent dull aching pain in the left flank for 3 months. History was unremarkable to suggest any urinary, virilising or feminising symptoms, hypertension and diabetes. The MRI of abdomen revealed a mass lesion measuring 7×5×5 cm with central area of necrosis arising from the antero -medial aspect of the upper pole of kidney. (Figure -1a, b)

Figure 1: MRI of abdomen showing a mass of size 7×5×5 cm with central area of necrosis arising from the antero -medial aspect of the upper pole of kidney. (Figure 1a – coronal section and 1b – axial section)

Figure 1a

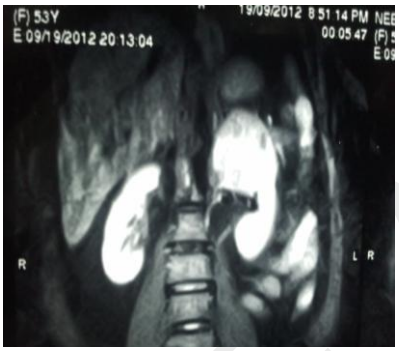
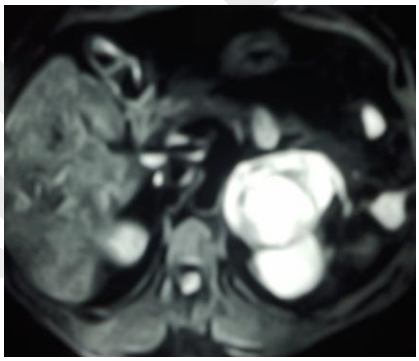


Figure 1b



The patient underwent tumor excision along with left sided nephrectomy. Histopathology revealed adrenal cortical carcinoma. (Figure - 2a, b)

Figure 2: Diffuse sheets of tumor cells with round vesicular nuclei and abundant eosinophilic cytoplasm.H&E x 20X (Figure 2a). Tumor showing large areas of necrosis and hemorrhage.H&E x 20X (Figure 2b).

Figure 2a

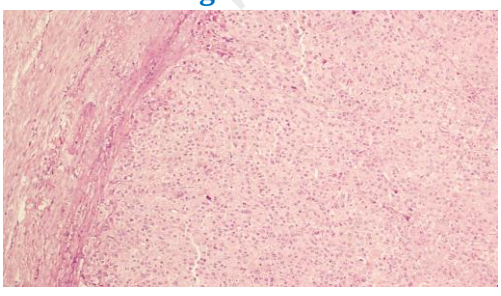
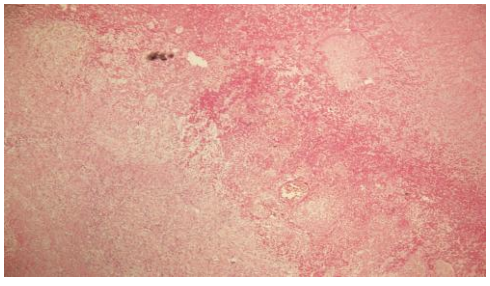
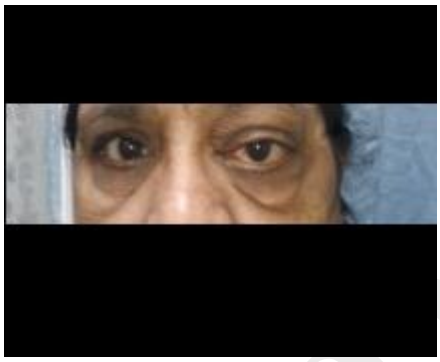


Figure 2b

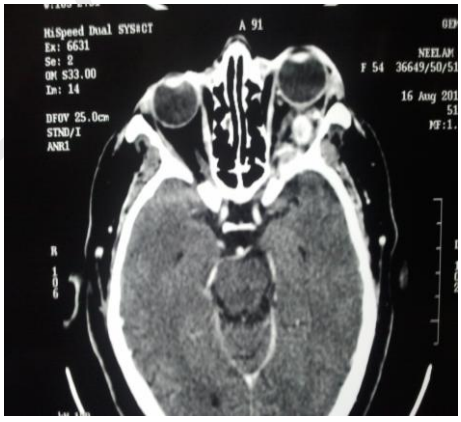


Patient did not take further treatment and presented after 1 year with protrusion of left eye. (Figure – 3) **Figure 3:** Photograph of a patient with proptosis of left eye



A contrast enhanced computed tomographic (CECT) scan of the orbit revealed multiple nodular lesions in intra and extra-conal compartments of left orbit with optic nerve compression. Bilateral orbital bones were normal. (Figure – 4) Based on these radiological findings, a diagnosis of orbital metastasis was established. CECT scans of abdomen & chest and bone scan were also normal. The laboratory studies did not show significant steroid hormone or catecholamine excess. Patient was subsequently planned for palliative radiation to the left eye and mitotane based chemotherapy.

Figure 4: CECT of orbit showing multiple soft tissue attenuation nodular mass lesions in intra-conal and extra-conal compartments of left orbit with optic nerve compression. Bilateral orbital bones were normal.



DISCUSSION

Adrenal cortical carcinoma is a rare tumour accounting for 0.05-0.2% of all malignancies [2]. There is bimodal age distribution with peaks in 1st and 4th to 5th decades of life. Slight female preponderance

is seen. Functional tumours occur in about 24-79% of cases. Majority of symptoms present are due to elevated levels of corticosteroid (30%-40%). Other hormones which can be elevated are androgen (20-30%), estrogen (6-10%) and aldosterone (2% to 2.5%). Around 30-40% patients have metastatic disease at initial presentation, while recurrent or metastatic disease develops subsequently in nearly all patients. [3] Common sites of metastasis are liver (48%-85%), lung (39%-60%), lymph nodes (7%-29%), and bone (7%-13%) [4]. Orbital metastases from ACC are rare, with a few cases reported in literature [5, 6, 7, 8]. In the majority, orbital metastasis was a component of widespread metastases from ACC. Isolated metastasis to orbit from ACC is even rarer. In 1932, Burch et al reported the 1st case of orbital metastasis from ACC from an autopsy series. Three probable cases and one possible case were described in a review of published reports. [5] In a survey of 1,376 orbital tumours from the Mayo Clinic, 1 case of orbital metastasis from ACC was reported in a 77-year-old woman. [6] Likewise, one patient with metastasis to the orbit has been listed among a series of 105 patients with ACC by Luton et al.[7] Another case of orbital metastasis was reported in a 24 year old patient of ACC by Bartley et al in 2001. The patient eventually died of widespread metastatic disease in bones, lungs and pelvis. [8]

The mainstay and the only curative treatment for ACC is surgical resection. However, the aggressive behaviour of this cancer limits the cure rate. The recurrence rate after complete resection is 35%-85% [1]. Mitotane based systemic therapy is used for treating inoperable and metastatic disease. Mitotane (1,1dichlorodiphenyldichloroethane), an analogue of a common pesticide, is an androlytic agent which promotes atrophy of adrenal tissue and is the only drug that has proven effective in treating patients with ACC. The response to mitotane is related to the drug's serum level.[9] The adverse effects of mitotane are often intolerable and patients rarely tolerate doses > 6 g/day for long-term therapy. The adverse effects of mitotane include confusion, somnolence, vertigo, ataxia, depression, weakness, headache, nausea, vomiting and diarrhoea. A low-dose mitotane regimen should be given with monitoring of the serum level of the drug to obtain an optimal effect. Systemic chemotherapy may be administered or combined with mitotane if the tumor is

poorly responsive to mitotane or the patient cannot tolerate the drug. Chemotherapy drugs effective in ACC are cisplatin, adriamycin and etoposide, but generally have disappointing results. Role of radiotherapy in ACC is of not much value and is reserved for the treatment of metastatic disease only. [10] The aim of treatment for orbital metastases is to improve the patient's quality of life by restoring or preserving visual function. Careful consideration should be given to the patient's general state of health, life expectancy, and the effects of treatment. Treatment for orbital metastases is mostly palliative in the sense that the presence of such metastases suggests hematogenous spread. Radiotherapy is often used to palliate orbital metastases and the recommended dose is 20–40 Gy delivered in fractionated manner. Radiotherapy alleviates symptoms in 80% of cases and in some cases restores vision [11]. Inhibitors of adrenal function, including drugs such as ketoconazole, metyrapone, and aminoglutethimide, radiotherapy, and radiofrequency ablation have also been used palliative [12]. Prognosis and treatment outcome depends on the tumor stage with 5 year survival rates of 84%, 63%, 51% and 15% for stages I, II, III and IV respectively.[2] Present treatment modalities are often disappointing in the management of advanced cases of ACC, and further studies are needed at molecular level, so that new treatment options can be offered. More well-designed studies are needed to determine the effects of various treatments.

CONCLUSION: ACC is a rare tumour that infrequently spreads to the orbit. It is an aggressive tumour with poor long term survival. The current treatment protocols are disappointing and there is a need for better therapies. Studies are needed at molecular level to understand the pathogenesis of this aggressive disease, so that better drugs are available to control this disease.

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