A case of Parathyroid Adenoma presenting as Brown's Tumor/? GCT Lower end femur (R)

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Abstract

Introduction: 20 years old female presented with solitary swelling over lower end of right femur since 7 months. Swelling was slowly increasing in size. Externally there were no skin changes, nor changes of infiltration of soft tissue. Preliminary examination of the patient revealed solitary osteolytic tumour with thinning of cortices, on X-ray / MRI suggestive of GCT / Brown's tumor.

Since MRI report mentioned Brown tumor as Diff diagnosis, S PTH was evaluated. It was significantly raised (1052.30pg/dl) Thyroid scintigraphy scan revealed? Parathyroid adenoma.

Endocrinologist opinion suggested the lesion as, Primary Hyperparathyroidism, and the patient was advised for 4D CT neck to look for parathyroid adenoma. The possibility of MEN2 was kept in mind wherein, the patient will require monitoring of Thyroid, parathyroid hormones & adrenal hormones during excision of endocrine tumor.

Due to the financial constraints, the patient was referred to higher centre (KEM Mumbai) as per the advice of In-house Endocrinologist.

The case which was posted for excision of tumor, packing the cavity with bone grafts & cement, did require the excision of the endocrinal neoplasm. Orthopaedic intervention was not required.

Introduction

20 years old female presented with solitary swelling over lower end of right femur since 7 months. Swelling was slowly increasing in size. Externally there were no skin changes, nor changes of infiltration of soft tissue. Preliminary examination of the patient revealed solitary osteolytic tumour with thinning of cortices, on X-ray / MRI suggestive of GCT / Brown's tumor. There was no pathological fracture. Coincidently it was noticed on her x-rays that she had similar swelling in (R) tibia U/3 as well.

Preliminary hematological investigations were inconclusive. She had similar swelling on clavical 8-10 yrs back, which was treated at that time, the details of which are not available.

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Endocrinologist opinion was taken which suggested Primary Hyperparathyroidism, and advised for 4D CT neck to look for

presentation of MEN 2 wherein the patient will require monitoring of Thyroid, Parathyroid & adrenal hormones during & post op period of the surgery. The patient needs to be observed for 3 months after surgery. In all probabilities the osseous structures will remodel to normal. If the need arises, then after that active orthopaedic intervention may be required.

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Literature

MEN1, or Wermer's syndrome, is inherited as an autosomal dominant trait. This syndrome is characterized by

- I) neoplasia of the parathyroid glands,
- ii) enteropancreatic tumors,
- iii) anterior pituitary adenomas, and
- iv) other neuroendocrine tumors with variable penetrance.

Other less common manifestations: foregut carcinoid, pheochromocytoma, subcutaneous or visceral lipomas

The estimated prevalence rate is 2–20 per 100,000 in the general population. This syndrome is caused by inactivating mutations of the tumor-suppressor gene MEN1 located at chromosome 11q13.

Treatment for most of these tumors, initial surgery is not

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curative and patients frequently require multiple surgical procedures and surgery on two or more endocrine glands during a lifetime.

MEN 2 consists of Medullary thyroid carcinoma (MTC) and hyperparathyroidism & pheochromocytoma. Three subvariants of MEN2A are

- I) familial medullary thyroid carcinoma (FMTC),
- ii) MEN2A with cutaneous lichen amyloidosis, and
- iii) MEN2A with Hirschsprung disease.

MEN2B is the combination of MTC, pheochromocytoma, mucosal neuromas, intestinal ganglioneuromatosis, and marfanoid features.

- I) Death from MTC can be prevented by early thyroidectomy. a RET proto-oncogene analysis should be performed on an individual with proven MEN2A.
- ii) Annual screening for pheochromocytoma in patients with germline RET mutations should be performed.
- iii) The goal is to identify a pheochromocytoma before it causes significant symptoms. The major question is whether to remove both adrenal glands or to remove only the affected adrenal at the time of primary surgery.
- iv) An alternative approach is to perform a cortical-sparing

adrenal ectomy, removing the pheochromocytoma and adrenal medulla, leaving the adrenal cortex behind.

Discussion

This case was misdiagnosed as GCT & we had actually planned for excisional biopsy with currattage & packing the cavity with bonegrafts & bone cement.

Since the tumor displayed multicentric foci & MRI revealed fluid levels, the second thought of some endocrine pathology, made us to explore further. The inhouse Radiologist mentioned in report of MRI the differential diagnosis as Brown tumor. Hence we decided to estimate S PTH which showed elevated levels (1152 pg/ml) Although the incidence of brown tumor is rare(1to7 in 1000), this suspicion has always to be kept in mind especially when the patient is young & the lesion is multifocal.

Conclusion

Before treating the local pathology in solitary lytic lesions of bone, it is essential to evaluate the endocrine status of the patient.

References

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Conflict of Interest: NIL Source of Support: NIL

How to Cite this Article

Verma P, Mulay S, Patil S. A case of Parathyroid Adenoma presenting as Brown's Tumor/? GCT Lower end femur $^{\circ}$. Journal of Trauma and Orthopaedic Surgery. Jan-March 2019;14(1):16-17.