

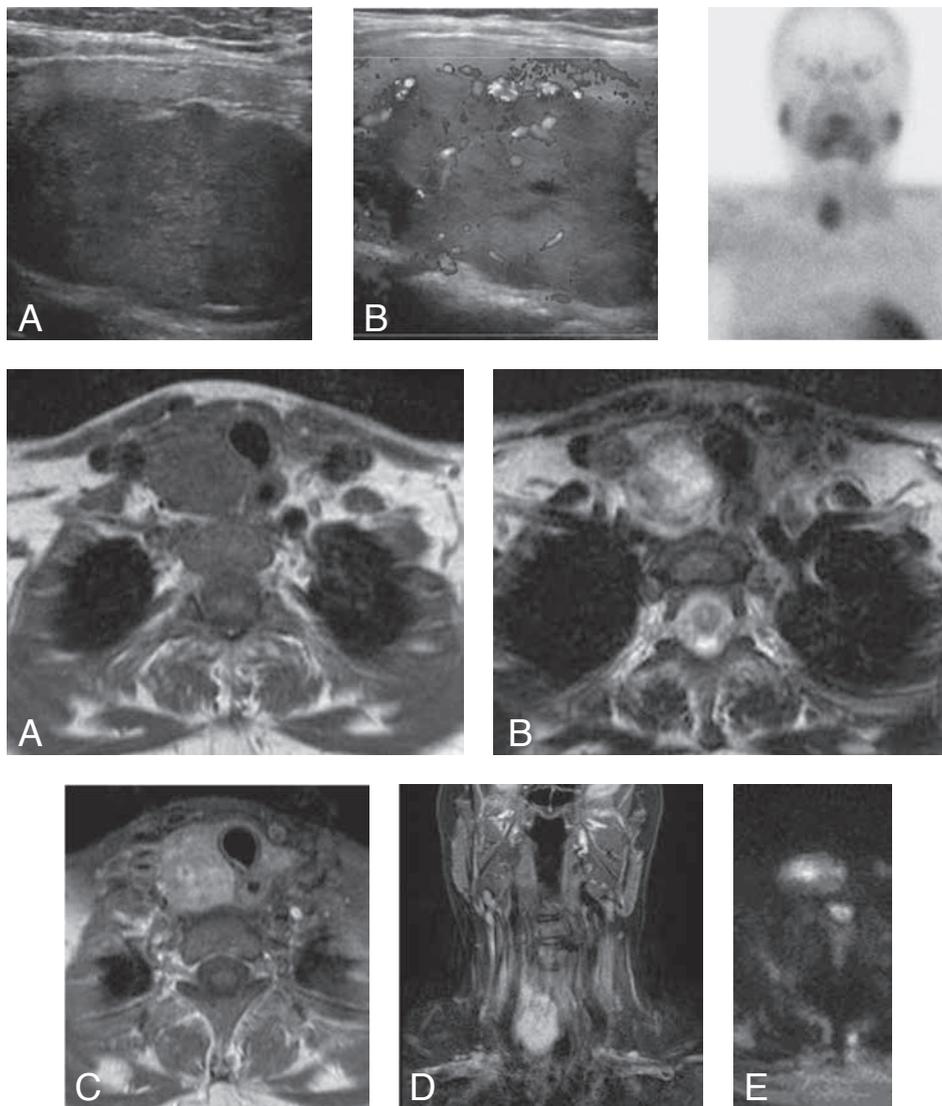
PARATHYROID CARCINOMA

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Key-word: Parathyroid, neoplasms

Background: A 32-year-old woman with palpable neck mass and clinical presentation of hypercalcemic crisis and primary hyperparathyroidism was referred to radiology and nuclear medicine departments for imaging studies.

The patient's medical history didn't reveal any findings related to neck irradiation, family history of parathyroid or calcium problems, nor any organ cancers. The laboratory tests showed severe hypercalcemia, a 7-fold increase in serum PTH concentration, high creatinine and normal phosphate serum levels.



	1A	1B	2
Fig.	3A	3B	
	3C	3D	3E

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Work-up

Ultrasonography and color Doppler ultrasonography of the neck (Fig. 1) shows a large (4.1 x 2.6 x 2.8 cm) hypo-echogenic solitary nodular lesion with slightly irregular borders localised in the right side of the neck is seen. On B, the mass shows increased peripheral blood flow.

On double-phase parathyroid scintigraphy (Fig. 2) two hours after intravenous injection of 20 mCi 99mTc-sestamibi, a persistent sestamibi uptake in the lesion of the right thyroid lobe is observed.

MRI of the neck area (Fig. 3, A: transverse T1-weighted image and B: T2-weighted image) demonstrates a mass localised in the right lobe of the thyroid gland presenting with low intensity on T1-weighted (A) and high-intensity on T2-weighted (B) images. On Gd-enhanced transverse T1-weighted axial image (C), moderate heterogeneous enhancement of the lesion is seen. On coronal STIR sequence image (D) and axial diffusion-weighted image (E), the mass shows high-intensity signal on the STIR sequence coronal image (D) and diffusion weighted sequence axial image (E).

Radiological diagnosis

Based on clinical and multimodality imaging findings obtained with ultrasonography, parathyroid scintigraphy and MRI of the neck, diagnosis of *parathyroid carcinoma* was proposed. This diagnosis was confirmed histopathologically after surgical resection of the lesion.

Discussion

Parathyroid carcinoma is a rare endocrine tumor and represents less than 1% of all cases with hyperparathyroidism.

The etiology of the disorder is unclear but a genetic predisposition (jaw tumor syndrome-germ line mutations in the gene CDC73) and a close relationship with some familial syndromes (familial isolated hyperparathyroidism, multiple endocrine neoplasia type 1 and type 2A) or irradiation of the neck have been reported.

An increased risk of atypia and carcinoma is reported for large (median size: 3.3 cm) hyperfunctioning parathyroid glands. A palpable neck mass can be shown in 22% up to 76% of patients with parathyroid carcinoma. In most cases, the dominant clinical picture is severe hypercalcemia. Severe nephrolithiasis, nephrocalcinosis, impaired renal function and bone disease are frequently seen. Surgery is the primary treatment of choice of parathyroid carcinoma. Although preoperative definitive diagnosis of parathyroid carcinoma is not

possible, preoperative and intraoperative decisions are very important to determine the extent of the surgical intervention and to achieve reasonable results. Long-term survival rates due to frozen section workup are not helpful.

The various imaging methods provide valuable information and contribute to the patient management by detecting, showing exact location and extent of the tumor, and by disclosing recurrence or metastases of the disease during preoperative or postoperative periods. The same imaging modalities are used to show both benign parathyroid tumors and parathyroid carcinomas. A combination of 99mTc sestamibi parathyroid scintigraphy and high-resolution ultrasonography offers high sensitivity and specificity (85% to 95%) for detecting of parathyroid pathologies. The most common cause for false-positive results is the presence of concomitant nodular thyroid disease. Parathyroid scintigraphy is used to define the primary lesion and to detect sites of recurrent and metastatic disease in functional parathyroid cancer, but it does not allow differentiation between benign and malignant tumors. Ultrasonography may show signs such as marginal irregularity, direct extension into adjacent structures and lymph node involvement, which are suggestive for malignant behaviour, but not always not specific. CT scan and MRI have initially been popular to show localisation of the primary parathyroid carcinoma. However, they may be more useful for detection of persistent, recurrent or metastatic foci in a wide range of regional or distant locations, including neck, lungs, bone and liver. These methods provide valuable information based on detailed anatomic information and perfusion characteristics. Finally, positron emission tomography (PET) has successfully been utilized to detect recurrent and metastatic parathyroid carcinoma. Although hybrid imaging modalities seem to be helpful in the evaluation of the metastatic disease from parathyroid cancer, it has been noted that brown tumors may sometimes lead to an incorrect interpretation on PET scan.

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