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Ectopic Parathyroid Adenoma in Child

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Abstract: A 10-year old girl presented with fatigue, hypercalcemia, and subperiosteal phalangeal osteolytic lesions. Ultrasonography and MIBI scintigraphy showed a structure near the lower pole of thyroid gland. The structure macroscopically appeared as adenoma, histologically it was thymic tissue. Bilateral neck exploration together with exploration of cervical thymic extensions was performed; adenoma was not found. During next two years, the level of calcium and parathormone raised, bone mineral density decreased. Ultrasonography, MRI, CT and PET/CT were negative. Adenoma was located by MIBI-SPECT/CT near the left border of jugulum. It was found dorsolateral to left common carotid artery and removed.

Introduction

Primary hyperparathyroidism is a generalized disorder of calcium, phosphate and bone metabolism, developing on the basis of long-term overproduction of parathormone with one or more pathologically changed parathyroid glands, on the basis of parathyroid adenoma, hyperplasia or, rarely, carcinoma. Primary hyperparathyroidism is relatively rare in children. When present, the multiple endocrine neoplasia (MEN) should be suspected. Solitary adenoma is a very rare event in children [1].

Case-report

A 10-year old girl presented in prae-collapse status. From three years of age, she had decreased appetite, significant fatigue, decreased muscle strength and slowly developing asthenic habitus. Physical examination of the child showed significant fatigue, otherwise, no particular signs were observed. Investigation revealed hypercalcaemia 2.89–3.03–2.76–3.11 mmol/l (norm 2.05–2.85 mmol/l), with increase in ionized calcium 1.5–1.59–1.6 mmol/l (norm 1.03–1.23 mmol/l), hyperparathoromonemia 12.0–5.0–8.1–11.0 pmol/l (norm 1.30–7.60 pmol/l) and increase in alkaline phosphatase 5.6 μkat/l (norm 0.8–2.3 μkat/l). The level of phosphate ions was normal. X-ray examination revealed subperiosteal brown tumor lesions in distal phalangs of both hands. MIBI scintigraphy was negative. Densitometry showed normal bone mineral density (0.827 g/cm^2). Neck sonography revealed a small hypoechogenic structure below the lower pole of the thyroid gland. Repeated MIBI scintigraphy suspected this structure to be the pathologically changed parathyroid gland. Standard bilateral neck exploration was performed. During the surgery, an isolated node, appearing macroscopically as a parathyroid adenoma was found. Peroperative histological examination but showed this was a thymic tissue. Thus, all typical and also ectopic locations of the parathyroid glands, together with the region of the cervical extensions of thymus were explored as described previously [2]. In addition, nodularly changed cervical extensions were removed and stored for subsequent detailed histological analysis. Pathologically changed parathyroid gland was not found, it was thus suspected to

be in the removed cervical extensions. Perioperative monitoring of parathormone levels was not reliable due to marginal levels of parathormone preoperatively. Sternotomy was not performed. Postoperative recovery of patient was without complications, the function of the recurrent laryngeal nerve was bilaterally normal, the wound healed normally. The calcium levels decreased to 1.80–2.80–2.67 mmol/l (norm 2.05–2.85 mmol/l), parathormone levels were 14.9–10.1 pmol/l (norm 1.30–7.60 pmol/l). Histologically, hyperplastic thymic tissue with frequent Hassall's corpuscles and also several lymph nodes was found. During the next 2.5 years, calcium level increased to 3 mmol/l, parathormone level to 19.44 pmol/l. Repeatedly, ultrasonography and MRI was performed with negative results. CT and PET/CT was also performed, again, with the same negative results. Bone mineral density rose to 0.839 g/cm^2. MIBI scintigraphy-SPECT/CT with the head bended backward finally revealed a suspicion on pathologically changed parathyroid gland near the left border of the jugulum. New MRI verified this localization; the structure was located dorsolateral to left common carotid artery, above the site where the left subclavian artery emerges from aorta. The reoperation was performed, when the patient was 13 years old. In described location, the parathyroid adenoma was found and removed. Postoperative recovery of patient was without complications, the function of the recurrent laryngeal nerve was bilaterally normal, the wound healed normally. Postoperative calcium levels normalized (2.36–2.37 mmol/l), the level of parathormone decreased to 7.94 pmol/l. The patient was repeatedly examined by endocrinologist. The fatigue had disappeared and muscle strength increased. Bone mineral density normalized.

Discussion

Primary hyperparathyroidism is not very common in children – they represent only 3% of all patients operated for primary hyperparathyroidism [3]. Clinical picture is not very specific in older children; often it begins with polyuria, polydipsia and obstipation. None of these was present in our case. Investigation often reveals hypercalcaemia, hypercalciuria, nephrolithiasis, bone mass changes and many non-specific signs such as fatigue, headaches, nausea, abdominal pain, diarrhoea, depression, or muscle pain [1, 3, 4]. Parathyroid adenoma occurs in 74–88% of cases in children older than 10 years [5]. Due to higher failure of primary operations in children (20% vs. 1% in adults), the combination of preoperative localization methods is absolutely necessary [3], as our case also indicates. The combination of MIBI and SPECT/CT might reach sensitivity up to 93% and specificity up to 95% [6]. The major causes of failure of the primary hyperparathyroidectomy are the presence of multiple pathological parathyroid glands, typically in MEN I or II syndrome, and/or ectopic localization of the parathyroid gland [7], as was in our case. Hyperparathyroidectomy is but a curative treatment and has much higher success than conservative treatment [8].

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