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Crippling bone deformities and a palpable neck lump; rare presentation of a giant parathyroid adenoma

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Abstract

The most common cause of primary hyperparathyroidism is a solitary parathyroid adenoma. The different manifestations of the disease are due to excess parathyroid hormone (PTH) secretion. “Giant parathyroid adenomas” weigh more than 2 g and are rarely evident in clinical examination. We present a young man who presented with severe bone deformities and a palpable neck lump. After surgery, a giant parathyroid adenoma was removed that weighed 33 g. Adenomas weighing more than 30 g are rare and presentation as a neck lump and severe bone disease has seldom been described. In patients presenting with a palpable neck lump, severe bone disease and renal calculi giant parathyroid adenomas should be considered as a possible culprit.

Keywords: Primary hyperparathyroidism, Giant parathyroid adenoma, Osteitis fibrosa cystica, Parathormone

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Introduction

The incidence of primary hyperparathyroidism is 25 cases per 100 000 per year (1). The most common cause in 85% of patients is a solitary parathyroid adenoma. Multiple adenomas, parathyroid hyperplasia and carcinoma account for the remaining cases (2). Parathyroid adenomas generally remain small in size and their average weight is between 70 mg to 1 gm (3). Those weighing more than 2 g are termed “giant parathyroid adenomas” (4). Interestingly, past studies revealed a direct relationship between gland weight and functional activity, as well as risk of post operative hypocalcaemia after parathyroidectomy (5,6). Generally, giant parathyroid adenomas do not produce severe symptomatic disease (7).

These tumours are not appreciable on clinical examination of the neck. Imaging modalities used for localizing the tumours are non-invasive. Ultrasonography (USG) of the neck, parathyroid scintigraphy and magnetic resonance imaging (MRI) of the neck may be used if surgical treatment is being planned (2).

We present the interesting case of a young man with severe recurrent bone and renal disease who presented with an enlarging neck lump. We were surprised to come across a giant parathyroid adenoma during surgery that was the culprit behind these features and was producing the swelling in the neck. This atypical scenario, of a giant parathyroid adenoma, manifesting as a neck lump and producing severe bone disease is a rare occurrence and deserves a

special mention.

Case Report

A-38-year old male labourer presented with progressively severe pain in his limbs and abdomen for the past 4 months. He had severe limb deformities including bowing of his lower limbs and kyphosis which had made him bed ridden. He also had severe epigastric distress which was diagnosed as acid peptic disease a few months ago. In addition he also had polyuria, polydipsia and weight loss. He described a small swelling in the neck for the last 6 months which had grown in size over the last 3 months. On examination, there was a palpable swelling in the lower right part of the neck which moved with swallowing. It was ovoid in shape, firm in consistency, non-tender with normal overlying skin and smooth surface, and had a size of 4×4×3 cm.

The thyroid hormone levels were normal. USG of the neck revealed a hypo echoic region in the inferior aspect of the right lobe of the thyroid gland, anatomically distinct from the gland.

Serum calcium level was 15.1 mg/dL. Serum parathyroid hormone (PTH) was 2500 ng/dL (normal 12-75 ng/dL) and alkaline phosphatase (ALP) was 3165 IU/L (44-147 IU/L). Serum phosphorus was 1.6 mg/dL (2.4-4.1 mg/dL). Serum urea was 52 mg/dL (20-40 mg/dL) and serum creatinine was 2.0 mg/dL (0.5-1.5 mg/dL)

X-rays of the long bones showed severe osteopenia and

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■ Implication for health policy/practice/research/medical education

In patients presenting with a palpable neck lump, severe bone disease and renal calculi giant parathyroid adenomas should be considered as a possible culprit.

lytic (punched out) lesions (Figure 1). X-rays and USG of the pelvis showed bilateral nephrolithiasis and severe osteopenia of the vertebral bodies (Figure 1). Technetium sestamibi scan revealed increased radio tracer uptake at the lower pole of the right thyroid lobe (Figure 2).

MRI of the neck revealed a 3.5×3 cm heterogeneous soft tissue intensity nodule in the posterior inferior aspect of the right thyroid lobe with anterior displacement of the thyroid suggestive of a parathyroid adenoma (Figure 3). Patient underwent a neck exploration for the adenoma, which was isolated from the right retro thyroid area (Figure 4). It was then sent for histopathology which revealed typical parathyroid tissue. Weight of the gland was 33 g with dimensions 5×4×4 cm. The PTH levels normal-

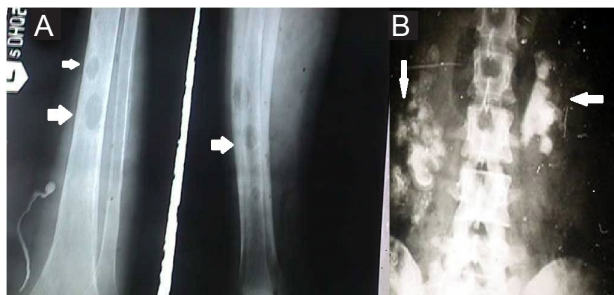


Figure 1. Panel A shows 2 views of the left lower limb. Notice the severe osteopenia, the lytic (punched out) lesions and the bowing of the bones in the second image of Panel A. Panel B is an x-ray of the pelvis showing large bilateral renal calculi and severe osteopenia of the vertebral bodies.

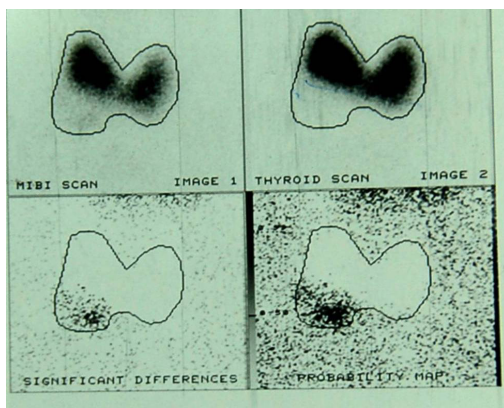


Figure 2. Technetium sestamibi scan of the thyroid gland; there is normal tracer uptake by the thyroid gland shown in the upper panels of the image. The lower panel shows an initial image showing increased uptake of the radio tracer in the lower pole of the right thyroid lobe. The delayed image shows persistent uptake of the tracer in the same area, suggestive of parathyroid adenoma.

ized 36 hours after surgery. There was no episodes of hypocalcemia and no intra or post procedure complication. Our patient’s symptoms resolved completely. He comes for routine follow up and is finally enjoying a healthy life.

Discussion

Giant parathyroid adenomas are an uncommon surgical disease (1). The primary mechanism for its clinical manifestations include increased production of the PTH culminating in hypercalcemia and hypophosphatemia by acting directly on the bones and kidney respectively, and on the intestines via vitamin D.

We searched the literature on the sizes reported and found only about 25 cases reported which weighed more than 30 g. The adenoma removed from our patient was 33 g. A direct relationship exists between the weight of the gland and its functional activity, however, severe bone disease has seldom been described with giant adenomas. The classic bone disease in hyperparathyroidism is called osteitis fibrosa cystica. This is diagnosed by the typical lytic or punched out lesions. There is accompanying severe osteopenia in the vicinity which makes bones prone to

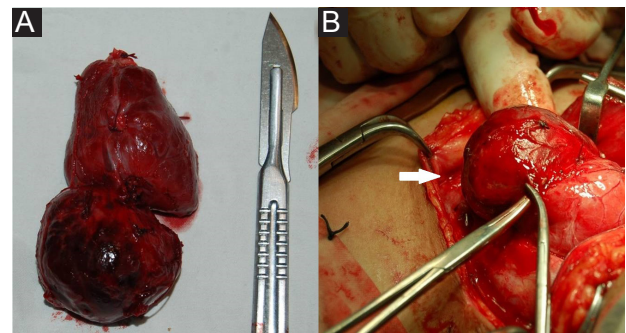


Figure 3. Magnetic resonance imaging of the neck revealed a 3.5 × 3 cm heterogeneous soft tissue intensity nodule in the posteroinferior aspect of the right thyroid lobe with anterior displacement of the thyroid suggestive of a parathyroid adenoma

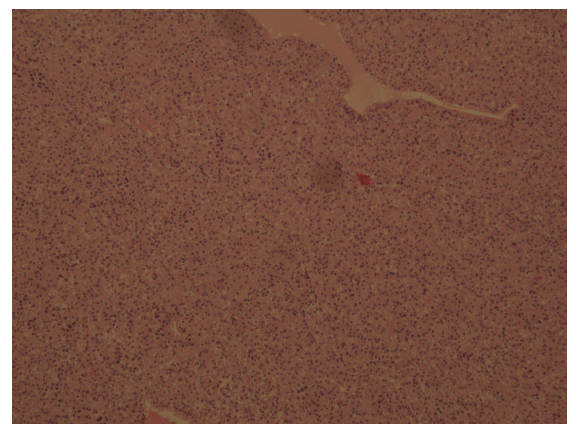


Figure 4. Post- and intraoperative views of the parathyroid adenoma. Panel A shows the large adenoma removed after the surgery. Panel B shows the adenoma after isolation during the surgery.

fractures and bowing. Our patient had severe parathyroid bone disease too (8-11).

The second rare aspect of our case apart from the size, was the presentation of the adenoma as a palpable neck lump. Parathyroid glands lie behind the thyroid glands and are not clinically palpable even when enlarged. Only a few cases reported, exist regarding this presentation. Imaging modalities further help in identifying the cause of the neck swelling as the parathyroid gland and aided in guiding us to the position of the gland and its estimated size. Parathyroid gland localization by various imaging modalities helps in decreasing operative time and potential morbidity. This also allows for a unilateral neck exploration to decrease risk of post operative hypocalcaemia and recurrent laryngeal nerve paralysis (9-12). Parathyroid scintigraphy accurately localizes the glands in over 80% of cases (13). Our patient's adenoma was localized accurately by this modality. USG of the neck complements sestamibi scan. This is most helpful in localizing intra-thyroidal parathyroids (9,13). In our patient this modality complemented sestamibi scan's findings. Computed tomography (CT) scan and MRI are less sensitive, however they may help in localizing mediastinal glands (13). The MRI scan is our patient, localized the lesion fairly well and gave us a good idea of the position of the gland pre operatively.

The most common approach to removing the adenoma is via a collar incision which also helps in examining all 4 glands for disease activity. A similar approach was used in our patient. Some investigations reported a higher risk of postoperative hypocalcemia and tetany in patients with giant adenomas. We monitored our patient stringently for tetany, however our patients did not experience tetany post-operatively.

Parathyroid adenomas produce an excess of PTH which causes hypercalcemia and hypophosphatemia. This is mediated by the PTH directly via its actions on the bones and kidney and indirectly through vitamin D in the intestines (2). Our patient had a typical biochemical picture of primary hyperparathyroidism corresponding to high calcium, low phosphorus and elevated PTH.

The most common presenting presentations of the disease include muscular weakness (70%), myalgia (54%), arthralgia (54%), constipation (32%) and nephrolithiasis (30%) and polyuria (28%). Less commonly a patient may present with symptomatic bone disease (2%).

The extreme bone deformities in our patient were due to a disease called osteitis fibrosa cystica as the classic bone disease in primary hyperparathyroidism. It presents with bony pains, fractures and bowing of weight bearing bones (8). Radiography of the long bones typically shows lytic or punched out lesions and severe osteopenia in the vicinity. The interesting aspect of our case was the discovery of a giant adenoma weighing 33 g that was palpable as a neck lump and produced this severe bone disease. Normally, each gland is 3 × 4 × 5 mm size and weighs between 70 mg to 1 g (1,4). Giant adenomas weigh above 2 g. Previous investigations revealed almost 25 case reports of giant adenomas weighing more the 30 g. The largest adenoma

removed was 145 g (9). Rarely, these adenomas may be bilateral (10).

Giant parathyroid adenomas usually do not produce such severe clinical disease and bone deformities, despite their large sizes and increased glandular activities. A direct relationship has been established between gland weight and its functional status (7). Only one case of giant non-functioning parathyroid adenoma has been reported (11). However, they may be associated with severe hypocalcemia post-surgery. Our patient however, did not develop hypocalcemia post-operatively.

Parathyroid gland localization by various imaging modalities helps in decreasing operative time and potential morbidity. This modality also allows for a unilateral neck exploration to decrease risk of post-operative hypocalcaemia and recurrent laryngeal nerve paralysis (12). Parathyroid scintigraphy accurately localizes the glands in over 80% of cases (13).

Conclusion

Through this case report, we concluded that giant parathyroid adenomas are an uncommonly encountered surgical disease and in very rare circumstances they may present as an enlarging neck lump and severe symptomatic bone disease.

Authors' contribution

All authors contributed equally to the manuscript.

Conflicts of interest

The authors declared no competing interests.

Ethical considerations

Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors.

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