CASE REPORT

Surgical management of acromegaly in a resource-challenged environment

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ABSTRACT

The management of acromegaly caused by an uncommon growth hormone-secreting pituitary adenoma can be challenging in low-resource African subregion. We conducted a study over a 2-year period to describe the results and challenges following surgical treatment of this rare condition in our centre. The clinical outcome was defined as successful based on the surgeon's intraoperative observation, postoperative neuroimaging findings and neuroendocrinological results. A total of three patients (two males and one female) aged 19-32 years were included. Visual impairment was the main presenting symptom in all the three patients. The postoperative period was uneventful. Acromegaly is an uncommon disorder in our region. Surgery is the treatment of choice in low-resource practice.

Key words: Acromegaly, growth hormone-secreting pituitary adenoma, insulin-like growth factor-I, pterional craniotomy

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INTRODUCTION

Growth hormone-secreting pituitary adenoma (GHPA) is a rare, chronic, systemic disease that is associated with premature death and significant morbidity.[1] It is related to high levels of growth hormone (GH) and insulin-like growth factor-I (IGF-I).

The standardised mortality index (the ratio of observed mortality in the acromegalic population to expected mortality in the general population) ranged from 1.2 to 3.3. If left untreated, patients with acromegaly can die approximately10 years earlier than the healthy subjects. According to prior studies, approximately 60, 25 and 15% of the patients die from cardiovascular disease, respiratory complications and cancer, respectively.[2,3] High GH/IGF-I levels and heart disease are the main factors related to poor outcome in these patients.[4] Some studies have shown that cerebrovascular disorders are a frequent cause of death, particularly among women, but they also involve patients who have been treated differently many years ago (craniotomy, radiotherapy);thus, a deleterious effect of these treatments (especially radiotherapy) cannot be ruled out.[1] Although the quality of life is affected in patients with GHPA, it can be partially improved by effective treatment.[5]

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Treatment options for acromegaly include surgery, medical therapy and radiotherapy. The cost of treatment including medications and the possibility of major side effects represent important limitations of the medical therapy.[2,6] In our region, a patient with acromegaly will need an estimated USD15,000 annually for medical management using octreotide. This is an astronomical amount compared with an operative option costing USD 2,000.

Surgical treatment provides rapid control of GH/IGF-I levels and is the first line of treatment for GH-secreting adenomas according to different neuroendocrinology societies and pituitary centres.[6,7] Classically, transsphenoidal microsurgery has been considered the best surgical approach for most GH-secreting adenomas. However, in the past decade, the endoscopic approach to the treatment of sellar lesions has become an important option for the resection of pituitary adenomas.[6-8] Some of the advantages of this approach include improved visualisation, less nasal trauma, increased patient comfort and, potentially, better results with respect to total tumour resection.[8-10]

In this case series, we aim to describe the results of surgery for the treatment of this rare condition in our centre. We also analyse the current literature related to the treatment of acromegaly and highlight the challenges encountered during the management.

CASE REPORT

The study included three patients (two males and one female) with age range of 19-32 years [Table 1]. Two patients were university undergraduates and one was a casual labourer. Visual impairment was observed in all the three patients [Figure 1]. The female patient had no light perception in the left eye and was also significantly worried about her amenorrhoea. The last two patients were concerned about their facial disfigurement. All patients had frontal bossing, skin thickening, painful joints, prognathism, jaw malocclusion, teeth separation and tongue and acral enlargement. Radiological evaluation was carried out [Table 2]. All the three patients had endocrinological features of pituitary stalk syndrome. The second and third patients had oestrogen and testosterone deficiencies, respectively. Electrocardiogram of the second patient revealed left ventricular hypertrophy. None of the patients had diabetes or hypertension.

The first patient initially declined surgery and opted for octreotide [Figure 2]. He was treated with short-acting octreotide (the only form available, 50 mg b.i.d.) and showed significant reduced joint pain. However, he was unable to continue the drug after only1 week of medication because of its high cost; he subsequently opted for surgery. The second patient initially declined surgery for spiritual treatment. However, her vision deteriorated and she opted for surgery [Figure 3].

Two of the patients underwent pterional craniotomy *ab initio*, and one abandoned the proposed endoscopic transsphenoidal approach and opted for craniotomy because of intraoperative instrumentation issues. All the patients had pterional craniotomy and total transcranial microscopic tumour excision. Gross total tumour resection was achieved in all cases with no morbidity. The postoperative period was uneventful, and the histology report confirmed pituitary adenoma. Postoperative IGF-I was normal 3 months after surgery in all patients [Table 1].

DISCUSSION

The most common cause of excessive GH secretion is a monoclonal benign pituitary tumour (adenoma) in more than 90% of the cases. This uncommon disorder is characterised by an acquired progressive somatic disfigurement, involving mainly the face and extremities, but other organs as well.

The disease affects both men and women equally,[11-13] with a prevalence of 40-70 cases per million inhabitants and an annual incidence of 3 to 4 new cases per million inhabitants.[3] We treated only 3 patients over a 2-year period. A recent study from Belgium suggests the prevalence of acromegaly to be approximately100-130 cases per million inhabitants.[14] In Germany, where screening of GHPA was performed by systematic IGF-I measurement, the prevalence of biochemical acromegaly

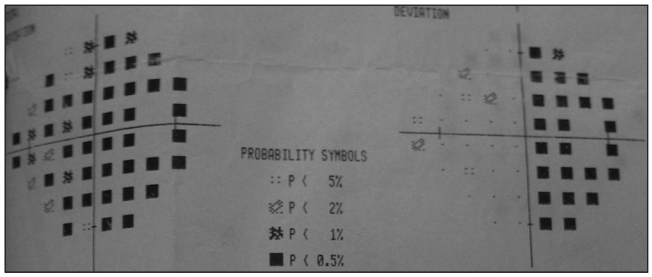

Figure 1: Automated perimetry of patient 3

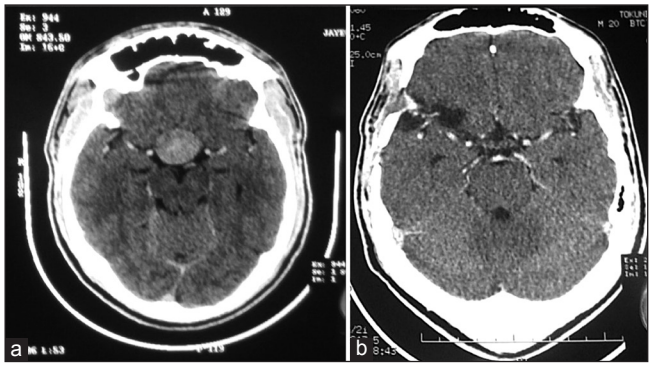

Figure 2: (a and b) Preoperative and postoperative contrast cranial computerised tomography of the first patient

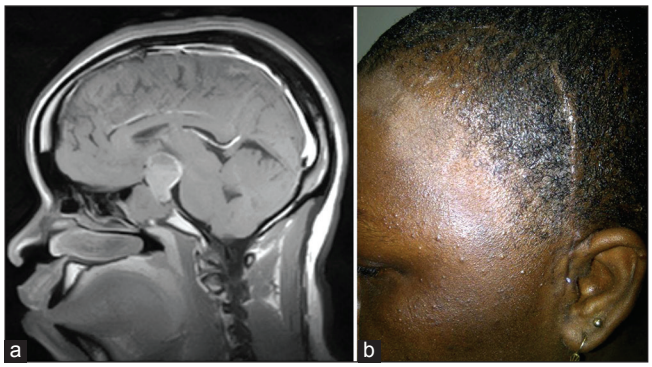

Figure 3: (a and b) Preoperative magnetic resonance imaging (MRI) and postoperative healed scar of the second patient

Table 1: Summary of the the patients demographics and results

Nos	Gender	Age (years)	Occupation	Vision	BMI (kg/m²)	Fasting GH (ng/ml)	Preoperative IGF-1 (ng/dl)	3 month spost operative IGF-1 (ng/dl)
1	Male	20	Undergraduate	Bitemporal hemianopia	31	20	650	110
2	Female	24	Undergraduate	No light perception in the left eye	39	21	686	269
3	Male	32	Labourer	Bitemporal hemianopia	38	35	720	140

Table 2: Radiological characteristics of the adenomas

Characteristic	No. of Cases
Tumour size	
Microadenoma	0
Macroadenoma	3
Suprasellar/parasellar extension	
Class A	0
Class B	0
Class C	1
Class D	2
Sella floor erosion	
Class I	0
Class II	2
Class III	1
Class IV	0
Cystic component	
Not present	2
Present	1

was even higher (1,043 per million).[15] Owing to its insidious onset, GHPA is often diagnosed late (4 to >10 years after onset), at an average age of approximately 40 years. The reasons for the rare occurrence of this condition in our centre may be due to lack of diagnosis, as many patients attribute their illness to spiritual forces and thus do not present to the hospital.

Symptoms and signs of GHPA are classical, but pre- and postoperative biochemical assays are not readily available in financially challenged regions. In addition, the cost of the test is quite prohibitive to most of the patients.

The specific goals of treatment are to reduce the volume of the pituitary tumour, avoid tumour relapse, normalise disease markers (GH and IGF-1), slow or reverse the clinical signs and symptoms, preserve normal pituitary function and restore life expectancy to the general population. Radiosurgery is not available in the country, and the use of medical treatment is not sustainable by most of our patients. The cost of medical treatments, which may be required indefinitely, must be weighed up against not only the risks of radiotherapy but also against the cost in our region. On the other hand, surgery is relatively more affordable and is probably, pragmatically, the best treatment of choice in low-resource practice. Globally, the use of preoperative treatment with somatostatin analogues is debatable.[5,16,17]

Surgery is recommended as the primary modality of therapy because it treats the local population with a good probability of a cure. Although one of the cases done in our study of a transsphenoidal approach had to be abandoned intraoperatively because of instrumentation issues (this is not an uncommon event though), surgery was successful in all cases with no morbidity.

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