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Case Report

Symptomatic Chiari type 1 malformation associated with acromegaly: A case report

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ABSTRACT

Background: Here, we report a patient who presented with both symptomatic acromegaly and symptomatic Chiari I malformation (CM1) with a C2-T5 syrinx.

Case Description: A 63-year-old female presented with bilateral arm dysesthesias and back pain. For approximately the past 30 years, she had chronic signs of acromegaly (i.e. an enlarged forehead, jaw, and nose, and enlarged hands and feet). When the cervical magnetic resonance showed a CM1 (tonsillar herniation) with C2-T5 syringomyelia, she underwent foramen magnum decompression and C1 posterior arch resection. Postoperatively, she was asymptomatic. The added finding of a growth hormone (GH)-producing pituitary lesion was treated medically with endocrine therapy, as she had incidentally required surgery/chemotherapy for a newly diagnosed colon cancer.

Conclusion: Symptomatic CM1, syrinx, and acromegaly may occur together. Appropriately treatment may include a suboccipital decompression, and C1 arch resection surgery, followed by either surgical or medical treatment for the GH-producing pituitary adenoma.

Keywords: Acromegaly, Chiari malformation, Pituitary adenoma, Syringomyelia

INTRODUCTION

A patient acutely presented with a Chiari I malformation (CM1), syrinx from C2-T5, and a pituitary adenoma (i.e. growth hormone [GH]-producing lesion resulting acromegaly). The former was managed with a suboccipital craniectomy and C1 laminectomy/fusion, while the accompanying pituitary lesion was managed medically/endocrinologically, due to the patient's newly discovered colon carcinoma warranting surgery, radiation, and chemotherapy.

CASE PRESENTATION

A 63-year-old female presented with 30 years duration of acromegaly (i.e. a large forehead, jaw, nose, hands, and feet), newly presented with bilateral arm dysesthesias, and neck pain. On neurological examination, she exhibited no focal deficits. The cervical magnetic resonance (MR) showed a CM1 (i.e. 7 mm herniation of the cerebellar tonsils of below the foramen magnum), a C2-T5 syrinx, while the brain MR documented a pituitary adenoma [Figures 1-3].

Table 1: Cases of combination Chiari I malformation and GH-producing pituitary adenoma.

No.	Authors	Cases		Symptom	Syringomyelia	Surgery	Progress after treatment
		Age	Sex				
1	Howell *et al*., 1924	38	Female	Headache Pain on hands		+ (FMD and for adenoma)	Improvement of headache
2	Macbride *et al*., 1925	59	Male	Headache Weakness			
		35	Female	Headache Pain on hands			
3	Lemar *et al*., 1994	29	Male	Cervical pain, paresthesias			
4	Hara *et al*., 1996	19	Female	Headache	+	+ (for adenoma)	Not listed
5	Agostinis *et al*., 2000	52	Male	Headache Hypoesthesia in the upper limbs	-	+ (for adenoma)	Reduction herniation
6	Ammerman *et al*., 2006	39	Female	Headache	+	+ (for adenoma)	Ascent of the cerebellar tonsils and resolution of the syrinx
7	Manara *et al*., 2013	-	Male	Hypoesthesia in the upper limbs	+		
		-	Female	Hypoesthesia in the upper limbs	+		
8	Present case, 2021	63	Female	Bilateral arm dysesthesia, back pain.	+	+ (FMD)	Ascent of the cerebellar tonsils and resolution of the syrinx

FMD: Foramen magnum decompression

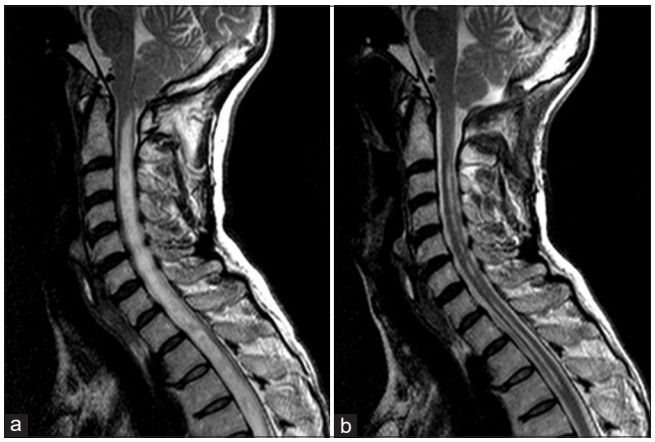

Figure 1: T2-weighted image on magnetic resonance imaging (a) is the preoperational image, which revealed cerebellar tonsil herniation and syringomyelia. (b) Syringomyelia was improved after the foreman magnum and C-1 decompression on image.

Endocrine evaluation

The preoperative endocrine examination revealed that GH and insulin-like growth factor-1 (IGF-1) levels were elevated at 13.8–369 ng/ml, respectively. Further, the GH level was not suppressed after the oral glucose tolerance testing.

Surgery and postoperative course

The patient underwent a suboccipital/foramen magnum decompression (FMD), with C1 posterior arch resection.

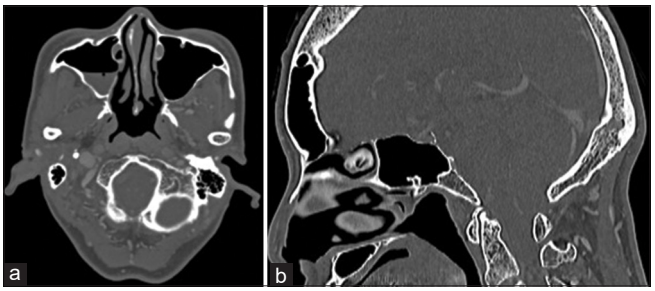

Figure 2: The axial image on computed tomography (CT) (a) showed that the transverse foramen magnum diameter was smaller than the anteroposterior diameter. The sagittal image on CT (b) showed the sagittal area on the midline posterior fossa.

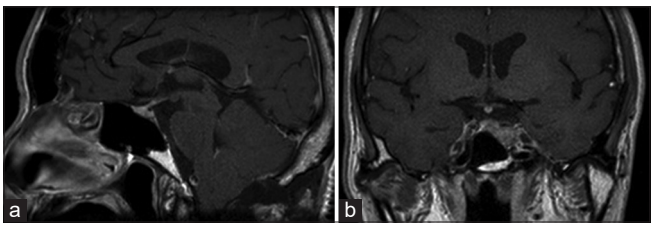

Figure 3: (a & b) T1-weighted image on magnetic resonance imaging showed pituitary adenoma.

Postoperatively, her forearm dysesthesias improved. Due to the need to treat newly diagnosed colon cancer, the patient's GH-producing pituitary lesion was managed medically (i.e. with a long-acting somatostatin analog therapy). The

subsequent endocrine examination after somatostatin analog therapy revealed that her GH and IGF-1 levels declined to 3.27–100 ng/ml, respectively.

DISCUSSION

In this case, the patient had a CM1, an accompanying C2-T5 syrinx, and a GH-producing pituitary adenoma. The authors found nine similar cases in of the literature; notably, these combined pathologies further contributed to posterior fossa bony hypertrophy and increased narrowing of the foramen magnum [Table 1].[1,2,4-8] Most cases with CM1 and acromegaly first received treatment for the pituitary lesion.[3] However, in our case, the patient first required a posterior fossa decompression/C1 laminectomy followed by medical management of the pituitary lesion.

CONCLUSION

Here, we presented a patient with a symptomatic CM1, syringomyelia, and a GH-producing pituitary adenoma (i.e. acromegaly). The former was effectively managed with a suboccipital/FMD, and C1 laminectomy, while the latter was treated with an endocrine therapy.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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