

Empty Sella Syndrome and Nonpuerperal Galactorrhea: A Case Report

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Abstract

Galactorrhea secondary to hyperprolactinemia is usually associated with a pathological or drug induced cause. We describe a 39 year old female patient who presented with a history of galactorrhea and no other specific symptoms. She had been on oral contraception for 15 years after the birth of her first child and had developed the milky nipple discharge three years prior to presentation. She had initially been managed as hyperprolactinemia secondary to long term contraceptive use but was referred for further investigation as she had no resolution to the galactorrhea after cessation of the drug for a significant period. Her blood investigations revealed an isolated persistently elevated serum prolactin and the diagnosis of an empty sella was made on radiological imaging.

Keywords: Galactorrhea • Hyperprolactinemia • Bromocriptine

Introduction

Galactorrhea presents as bilateral breast-milk production unrelated to lactation. It may occur in patients up to one year post pregnancy; those who have discontinued breast feeding; nulliparous or post-menopausal women as well as in male patients. In newborns, a high level of estrogen from the mother may cause transient galactorrhea termed 'witch's milk' [1].

It is often the result of hyperprolactinemia secondary to lactotroph adenomas, hypothyroidism, chronic renal failure, cirrhosis, chest wall burns, breast augmentation mammoplasty, polycystic ovary syndrome or pharmacological interruption of the hypothalamic- pituitary dopaminergic pathways [2].

Medications commonly associated with galactorrhea are selective serotonin reuptake inhibitors, gastrointestinal motility drugs and antipsychotics [3,4].

Empty sella syndrome has often been associated with hyperprolactinemia and galactorrhea in the absence of any primary pituitary pathology [5]. It typically presents with ocular or intracranial symptomatology and is frequently an incidental finding on patients who present for investigation of other pathology [6]. Empty sella syndrome occurs when the subarachnoid space herniates into the saddle filling it with Cerebrospinal Fluid (CSF) while flattening normal pituitary tissue and elongating the stalk. It is termed primary empty sella when there is no known pathological or iatrogenic cause.

One theory suggests that during pregnancy, with the increased hormonal demand the pituitary gland may enlarge significantly and thereafter retract post-delivery, leaving a larger space in the pituitary fossa for herniation of the subarachnoid fluid to occur [7].

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Case Presentation

A 39 year old female with a 3 year history of bilateral breast milk production, presented to the surgical department for investigation of breast pathology as a cause for her galactorrhea. Initially attributed to her chronic use of oral contraception for 15 years prior, she had been placed on bromocriptine and the contraceptive drugs stopped. She had no resolution of the galactorrhea despite treatment and no visual field or neurological symptoms.

On clinical examination she was haemodynamically stable with a Glasgow Coma Scale of 15 and a normal general exam with no neck masses or features of chronic liver disease noted. Eye examination revealed no visual field abnormality and she exhibited a normal neurological exam. Breast examination demonstrated the bilateral milky nipple discharge with no associated breast masses.

Blood investigations displayed an isolated hyperprolactinemia of 109 ug/L and normal thyroid, liver and renal function tests. The hyperprolactinemia was noted to be persistent when previous blood investigations were reviewed.

Radiological investigations revealed a normal mammogram with no breast pathology noted and Magnetic Resonance Imaging (MRI) of the brain demonstrated characteristics of an empty sella with no features of raised intracranial pressure (Figures 1-3). She opted for conservative treatment with cabergoline. On 3 month follow up her galactorrhea had largely resolved.

Results and Discussion

The aetiology of galactorrhea in premenopausal women is more often physiologic in nature and initial evaluation must exclude pregnancy and breastfeeding within the past year. When associated with amenorrhea it is often caused by hyperprolactinemia [4,8].

For secondary causes a thorough pharmacological history must be obtained. Clinical examination should include a comprehensive ocular and neurological exam [2,4,8]. A serum prolactin measurement must be obtained, this normally rises up to 500 ng per mL during pregnancy. Levels below 100 ng per mL are more often associated with drug-induced hyperprolactinemia and levels above 250 ng per mL may suggest a pituitary adenoma [8]. Other investigations include thyroid, liver and renal function testing to rule out other causes [2-4].

Radiological assessment of the pituitary is indicated once endocrine and systemic causes have been ruled out. Computer Tomography (CT) with



Figure 1. MRI T1 sagittal view demonstrating an elongated pituitary stalk, CSF filled pituitary fossa with a barely perceptible flattened pituitary gland.

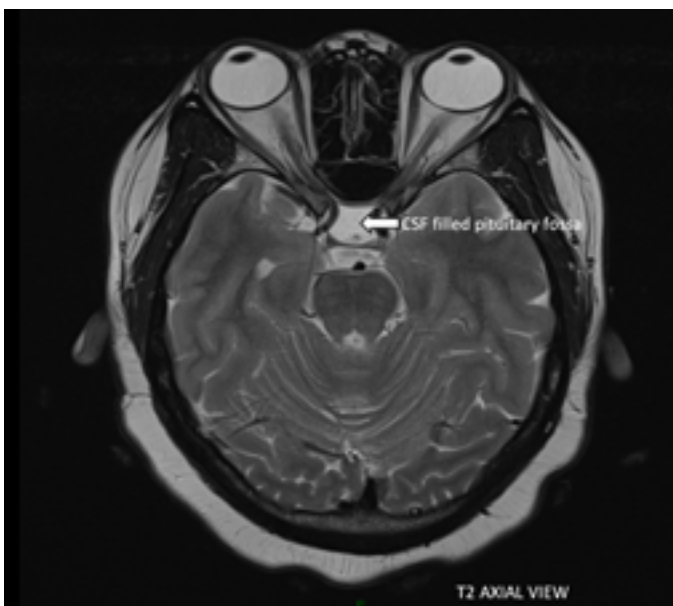


Figure 2. T2 axial demonstrating the central pituitary stalk and a CSF filled pituitary fossa.

contrast may be done however they may not be sensitive enough to identify small lesions or large lesions that are isodense with surrounding structures [6,7].

MRI is the preferred modality for confirming the diagnosis. Typical imaging findings include a sella filled with CSF, a flattened and barely visible pituitary gland and the infundibulum may be seen traversing the space to lie in its normal central position, thereby excluding a cystic mass. This is known as the infundibulum sign. The differential diagnosis may include other cystic lesions of the pituitary gland, all of which displace the infundibulum to the side of the fossa (i.e. absent infundibulum sign) [9].

If orbital findings such as optic nerve sheath prominence and fattened posterior globes are present, particularly in middle aged female patients, undiagnosed intracranial hypertension may also be considered in the differential.

Other radiological investigations may include imaging of the breasts to rule

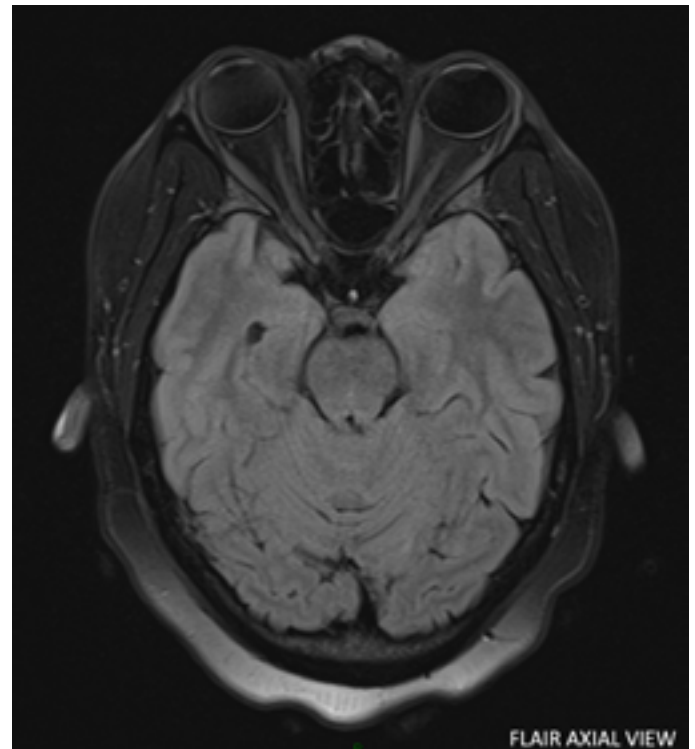


Figure 3. MRI axial FLAIR also demonstrating CSF filled pituitary fossa.

out local pathology and pelvic ultrasound where polycystic ovary syndrome is suspected.

Management is typically with a dopamine agonist, usually bromocriptine as it has been used for the longest period and the associated side effects are well documented. These include gastrointestinal upset, headaches and dizziness. Cabergoline is used in cases where patients are either resistant to or intolerant of bromocriptine. Another drug used in resistant cases as it has high pituitary selectivity is Quinagolide, the disadvantage of this drug is that it has a limited daily use [2,8].

Conclusion

Galactorrhea and amenorrhea is frequently associated with hyperprolactinemia secondary to a multitude of causes, the most common being pharmacological. Patients with no other symptomatology should be considered for radiological investigation if galactorrhea persists despite cessation of causative medications and dopamine-agonist therapy.

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Ethics Approval and Consent to Participate

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Patient Consent for Publication

Patient signed consent form complete.

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