Isolated Intrasellar Pituitary Mucocele

İzole İntrasellar Hipofiz Mukoseli

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ABSTRACT

Mucous cyst or mucocele are epithelial lined cavities containing mucous that occurs mainly in the frontal and ethmoid sinuses. Intracranial mucocele is a rare occurrence. We are presenting an interesting case of a young lady, presented with fainting spells, syncopal attack, recurrent frontal headache and blurring of vision for at least 2 months prior to presentation to us. CT scan showed shows a hyperdense mass arising from the tuberculum sellae with no foci of calcification within and no erosion of the adjacent bony structures. The intrasellar mucocele was diagnosed following surgery via trans-sphenoidal approach and biopsy with confirmatory histopathology.

Keywords: Mucocele, Pituitary, Cranial

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ÖZET

Mukoza kisti veya mukosel esas olarak frontal ve etmoid sinüslerde meydana gelen mukus içeren epitel ile kaplı boşluklardır. İntrakraniyal mukosel nadir görülen bir durumdur. Bize başvurmadan en az 2 ay önce bayılma nöbetleri, senkop atağı, tekrarlayan frontal baş ağrısı ve görme bulanıklığı ile başvuran genç bir bayanın ilginç bir olgusunu sunuyoruz. BT taraması, içinde kalsifikasyon odakları olmayan ve bitişik kemik yapılarında erozyon olmayan tüberkülum selladan kaynaklanan hiperdens bir kitle gösterdi. İntrasellar mukosel tanısı trans-sfenoidal yaklaşım ve doğrulayıcı histopatoloji ile biyopsi ile ameliyat sonrası konuldu.

Anahtar Sözcükler: Mukosel, Hipofiz, Kranial

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INTRODUCTION

A mucocele is a slowly expanding epithelial-lined lesion filled with aseptic mucus(1). Its expansion can lead to thinning, distension and erosion of adjacent bone. It occurs as a result of blockage to the sinus ostium due to congenital cause or as a result of trauma, surgery, infection, anatomical abnormality, amongst others(1,2).

Mucoceles primarily occur in the frontal sinuses (60% to 65%) but may also be found in ethmoid sinuses (20% to 25%), maxillas (10%) and sphenoid sinuses (1% to 2%)(1,3). Intracranial mucocele is extremely rare. Mucoceles prevalence is higher between the third and fourth decades of life with slight male predominance(1,2).

Symptoms are dependent on the location of the mucocele and the degree of expansion into surrounding structures. Patients may present with symptoms which includes, headache, facial pain, facial edema, nasal obstruction, diplopia, decreased visual acuity, epiphora, meningitis or sign of central nervous symptoms inflammation(1,2).

Both Computed tomography (CT) and magnetic resonance imaging (MRI) can be utilized to aid in diagnosis. CT is sensitive in delineating bone erosions, while MRI had the advantage of multiplanar imaging and was much more sensitive for differentiating mucocele from a tumor on the basis of high signal intensity characteristics in MRI. Signal intensity in T1 weighted images is determined by mucus viscosity and protein concentration while signal intensity in T2 weighted images is determined by the water content(1-3,7).

In the past, external approaches were employed, such as Lynch-Howarth incision, Caldwell-Luc approach or an osteoplastic flap. Currently, the trend has shift to endoscopic approach as it has shown to reduced morbidity and recurrence rate(2). The aim of the surgery is diagnosis, drainage of the content and excision of the cyst wall(3). Complication of the operation are CSF rhinorrhea, pneumocephalus and infection(4). We performed minimally invasive approach namely transsphenoidal resection in managing intracranial mucocele for this patient.

CASE REPORT

An 18 years old woman was admitted to our facility presenting with fainting spells. Her past medical history was positive for rhinitis and she also have allergies to dust. There was no history of head trauma or previous surgery. She complains of recurrent syncope attack preceded with blurring of vision over 2 months. She also complains of mild intermittent frontal headache.

Physical examination was unremarkable with no neurological abnormality. Nasoendoscopy examination revealed bilateral inferior turbinates hypertrophy and congestion with no nasal discharge and high deviated nasal septum on the left side. Visual assessment was normal. No cardiac arrhythmias detected in Holter monitoring.

Laboratory test including complete blood count test and hormonal studies (Cortisol, FSH, Estradiol and Prolactin) were within the normal range.

Computed tomographic (CT) scan in Figure 1 below shows a hyperdense mass arising from the tuberculum sellae with no foci of calcification within and no erosion of the adjacent bony structures. Magnetic resonance imaging (MRI) shows thin walled cystic cellar lesion which shows high T1 weighted and low T2 weighted signal measuring 16mm x 19mm x 23mm (AP x W x CC) with suprasellar extension causing compression of the optic chiasm. No enhancement demonstrated post contrast. Mild mucosal thickening in bilateral maxillary sinuses noted.

Patient underwent endoscopic resection of the posterior nasal septum and a trans-sphenoidal approach was made through the sphenoid sinus to the sella turcica. The tumor visualized at the suprasellar region is firm with calcifications at few regions with some mucoid discharge. Complete removal of the tumor was achieved. Post-operative recovery was uneventful, and patient were discharge well on day 3 post operation. Upon review in clinic, patient was completely well, no active issues reported and all the symptoms before operation disappeared.

Analysis of the biopsy specimens revealed features diagnostic of mucocele. Macroscopically, histopathological examination shows fragments of whitish grayish tissue. Microscopically, as depicted in figure 2 below, shows sections of eosinophilic mucous like materials with few inflammatory cells and calcification and no evidence of epithelial lining.

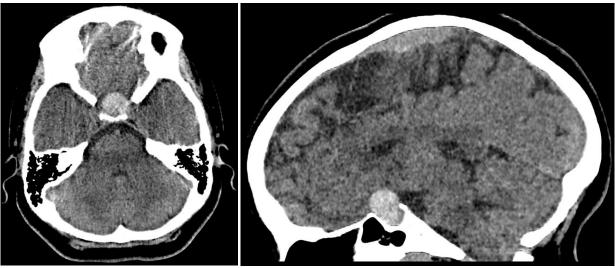


Figure 1. Axial and sagittal plain CT Brain images showing a well-defined intrasellar hyperdense lesion with suprasellar extension.

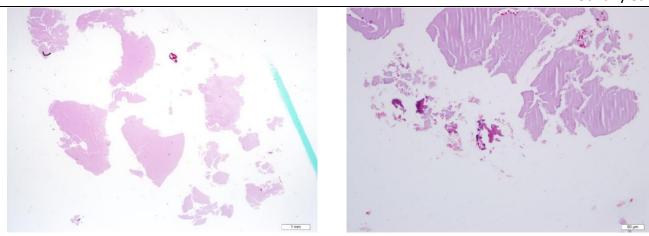


Figure 2. Eosinophilic mucous like materials with few inflammatory cells and calcification.



Figure 3. Axial and sagittal MRI Brain images showing a well-defined intrasenar hyperuense lesion with suprasenar extension reducing in size post-operation

DISCUSSION

A mucocele is rarely found in the intrasellar region, although mucoceles of other paranasal sinuses are not uncommon. The sella turcica is a depression in the body of the sphenoid bone in which the deepest part known as the hypophyseal fossa holds the pituitary gland. The pathogenesis of isolated intrasellar mucocele is debatable as the normal pituitary gland does not have any mucus secreting constituents within it(5). The sphenoid sinus is not limited to the body of the sphenoid bone as extension of the air-filled sinus into other parts of the sphenoid bone occurs occasionally(6).

Pneumatized recesses are often found in the greater wing and base of pterygoid process. Infrequently, there are extension superiorly into the lesser wing or anterior and posterior extension into the basilar occipital bone(6). No case has been reported in the literature to date of an isolated intrasellar mucocele without any connections to the paranasal sinuses or without previous history of sinus surgical intervention. We have hypothesized that there can also be an inferior extension of the pneumatized recesses into the sella turcica in which precipitation of an inflammatory reaction could result in mucocele formation of the sella turcica.

There are also other hypotheses about their formation which includes obliteration of the pneumatizing air cell tracts or that such mucoceles develop from cell rest in the inner or under half of the sella turcica(3). There is also speculation that its origin was secondary to an aberrant sinus without an apparent ostium(3). Limited case of mucoceles involving the sella turcica have been reported. Most occurs as extension from the sphenoid region by means of bony erosion and some occurs as a sequel following a trans-sphenoidal sinus surgery. The latter can be due to implantation of the nasal mucosa during surgery which invades into the pituitary fossa(5).

CT is useful for detection of bony erosion and its extension and also to assess presence of sinus pathology. Generally, mucocele is isodense to the brain parenchyma, with attenuation values varies from 10 to 40 HU which reflects the hydration and protein contents of its mucus. The higher its attenuation value, the older the lesion is. Fine calcifications may be demonstrated within the mucocele which represents an inspissated dehydrated mucocele content. MRI has the advantages of multiplanar imaging in to differentiate the lesion from other soft tissue tumors on the basis of MR signal intensity characteristic. Mucocele signal intensity in T1 weighted and T2 weighted images varies, depending on its content with no contrast enhancement. Except for a regular linear subtle peripheral enhancement on T1 weighted images with increased protein content. Initially, the contents will be predominantly aqueous, so the corresponding image will be hypointense on T1 weighted and hyperintense on T2 weighted. As mucocele ages, the protein contents may increase with variable degree of hydration of the mucus resulting in a T1 weighted hyperintensity and T2 weighted hypointensity(7). Enhancement within the center of the lesion or nodular peripheral enhancement should raise the suspicion of a coexisting tumor(3,6,7).

In our case, the CT and MR images shows features of an old dehydrated and inspissated non-invasive mucocele with CT characteristic of a hyperdense lesion of 50-60 HU attenuation with no adjacent bony erosion and MR images of the lesion showing high T1 weighted and low T2 weighted signal with no enhancement post contrast.

The aim of surgery on a mucocele is complete resection of its lining. In this case, removal of the intrasellar tumor via trans-sphenoidal approach is preferred due to minimal trauma as it carries less risk than transcranial surgery. And this is to avoid intracranial contamination if there is ongoing infectious process. External approach is more aggressive with high morbidity and mortality and is used in extreme cases with significant tumor extension(3). In the future, more cases could be conducted and reviewed with similar way.

Minimally invasive approach has become the new methods in managing pituitary cases nowadays.

CONCLUSION

Isolated intrasellar pituitary mucocele is a rare entity and its etiology remains controversial. While mucocele may not be of immediate concern, evaluation is highly recommended as local extension can lead to compression of adjacent structures and also to rule out other condition that may have similar appearance. Surgical intervention which consists of drainage and removal of membrane lining via trans-sphenoidal approach remains as the treatment of choice in this case.

Conflict of interest

No conflict of interest was declared by the authors.

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