

Case Report

A Case of Giant Dermoid Cyst at Sacrococcygeal in Adult at "X" Hospital in Bandung

Laporan Kasus Giant Kista Dermoid pada Regio Sakrokoksigeal di Rumah Sakit "X" di Bandung

Farid Yudoyono^{1,2*}, Christian Ade², Hasan Baraqbah^{1,2}, Adhitya R Yudhadi¹

¹Neurospine and Pain Center, Santosa Hospital Bandung Kopo

Jl. K.H Wahid Hasyim No.461-463, Bandung, Jawa Barat, 40227, Indonesia

²Department of Neurosurgery, Padjadjaran University Faculty of Medicine, Bandung Indonesia

Jl. Pasteur no.38, Bandung, Jawa Barat, 40161, Indonesia

**Corresponding author*

Email: faridspine@gmail.com

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Abstract

Dermoid cysts are benign tumors that can be found anywhere in the body. There is no dermoid cysts clear data yet in Indonesia. In the spine, these tumors generally occur in the lumbar region and are often accompanied by congenital spinal dysrafism and/or dermal sinus passages. Dermoid cysts arise from abnormal growth of the ectodermal folds during pregnancy. Its incidence is 0.7 – 1.8% of all central nervous system tumors. The aim of reported this case is to highlight the role of a diagnostic and evaluate the surgical outcome. A 47 year old female patient with a lump in the sacrococcygeal area measuring 20 x 18 x 16 cm, with hard and soft consistency. MRI images show a hypo and hyperintense mass without spinal deformity. After the tumor was removed, the patient was allowed to go home for the 2nd day without complications. In conclusion, extraspinal dermoid cyst is a rare case that requires special diagnostic and surgical modalities to provide better clinical results.

Keywords: *dermoid cyst; excision; sacrococcygeal; spinal dysraphism*

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Abstrak

Kista dermoid adalah tumor jinak yang dapat ditemukan di mana saja di tubuh. Insiden kasus kista dermoid di Indonesia belum ada data yang pasti. Di tulang belakang, tumor ini umumnya terjadi di daerah pinggang dan sering disertai disrafisme tulang belakang bawaan dan/atau saluran sinus dermal. Kista dermoid timbul dari pertumbuhan abnormal lipatan ektodermal selama kehamilan. Insidensinya adalah 0,7 – 1,8% dari semua tumor sistem saraf pusat. Tujuan dari penulisan laporan kasus ini adalah untuk menekankan pentingnya peran diagnostik dan mengevaluasi hasil bedah. Seorang pasien wanita berusia 47 tahun dengan benjolan di daerah sacrococcygeal berukuran 20 x 18 x 16 cm, dengan konsistensi keras dan lunak. Gambar MRI menunjukkan massa hipo dan hiperintens tanpa kelainan bentuk tulang belakang. Setelah tumor diangkat, pasien diperbolehkan pulang untuk hari ke-2 tanpa komplikasi. Simpulan, kista dermoid ekstraspinial merupakan kasus langka yang memerlukan modalitas diagnostik dan bedah khusus untuk memberikan hasil klinis yang lebih baik.

Kata kunci: eksisi; kista dermoid; *spinal dysraphism*; sakrokoksigeal

Introduction

Dermoid cysts are benign tumors when located in the spine usually accompanied by congenital spinal dysraphism or the dermal sinus tract.^{1,2} Dermoid cysts originate from growth abnormalities in the ectodermal fold. The peak incidence of these diseases occurs in the age of twenty to thirty.³ The symptoms are caused by the effect of the increased volume of the tumors which has compression effect to the surrounding nerve and soft tissue.^{3,4} Rupture of the skin is associated with high rates of morbidity and mortality.⁵ Dermoid cysts are rare disease, where the disease only covers 0.7-1.8% of all central nervous system tumors, especially tumors that occur in adults and are located in the intramedullary.⁶

MRI is the choice of imaging modality for this case. These cysts generally have uni- or multilocular cyst lesions with varying intensity characteristics because of the varying cyst contents. Components of the cyst are divided into two: fluid and lipids. Fluid in cysts gives a picture of hypointense, whereas lipids in cysts give a picture of hyperintense on T2WI MRI. Based on the components that are most numerous in cysts, the intensity of MR images can be different. Generally, enhancements to intravenous contrast are low, depending on the amount of soft tissue in the cyst.^{7,8} Other abnormalities can occur together with dermoid cysts, for example myelomeningocele, hypertrichosis, dermal sinus tract, or a combination of all three.^{3,7} In dermoid cysts, skin components, such as oil glands, sweat glands, hair and hair follicles can be found; these skin components are not found in epidermoid cysts. Yellow cystic fluid generally contains desquamated epithelium and oil gland secretion products.⁷

This is slow-growing tumor, so the signs and symptoms of these tumors are found in the age of around twenty to thirties.³ and compress of nerve structures around the tumor. All

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symptoms of a large space-occupying-lesion in the spinal cord can be seen, but have different intensities. In some literature, the number of male sufferers is slightly more than females.^{3,4}

The risk of morbidity and mortality is associated with intact cystic walls. In the event of rupture, the prognosis is not good, where there is spread of lipid droplets in the spinal cord.⁵ Although passive transport of lipid droplets through the cerebrospinal fluid pathway is still unknown, dissemination of lipids to the spinal cord to the cistern and intracranial ventricles can occur through the central canal.⁷

Spontaneous, traumatic, or intraoperative rupture can be asymptomatic or can cause symptoms such as headaches, nausea, vomiting, meningism, or worsening neurological symptoms that can cause coma. These symptoms are thought to be symptoms of chemical or aseptic meningitis or secondary arachnoiditis caused by leakage of fatty and proteinous components in the subarachnoid space. Excision is the main treatment that can reduce symptoms.^{8,9}

Although dermoid cysts are mostly benign tumors, there are several case reports that describe the transformation of these tumors into malignant tumors in the form of squamous cell carcinoma. If dermoid cysts are excised completely, recurrence is rarely found in these cases. Previous study reported the dermoid cysts that turn into malignant tumors are generally associated with ovarian and intracerebral lesions.¹⁰⁻¹² Ino et al reported a case of a tumor found in the lumbar vertebra which then turned into squamous cell carcinoma 6 years after surgery.¹³ The etiology of the transformation to a malignant tumor is still unclear. It is concluded that the cyst was actually malignant, but spread slowly.¹⁴⁻¹⁶ This case is unique because dermoid cyst occurred in extraspinal space. The aim of reported this case is to highlight the role of a diagnostic and evaluate the surgical outcome

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This case report has received Ethical Clearance from Santosa Hospital Committee No 483a/KET/DIR/SHBK/XII/2020. Female, 47-year-old presented with the complaints of a mass in the sacrococcygeal region for 27 years and gradually increased in size. No complaints of lower limb weakness or bowel and bladder dysfunction. On physical examination, a giant mass 20 x 18 x 16 cm in size. It was of variable consistency. From the non-contrast lumbosacral T2WI MRI sagittal and axial show hypointense mixed with hyperintense and well demarcated. No bone defect. (Figure 1) The patient was performed excision with mass size mass size 20x18x16 cm solid and capsulated, patient was discharged on day 2 without complication. (Figure 2)

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Figure 1 A. T2WI MRI Sagittal Image; B. Axial Show Hypointense Mixed with Hyperintense and Well-Demarcated Lesion



Figure 2 A. Lateral View; B. Superior View Before Surgery; C. Macroscopic Mass After Surgery Size 20x18x16 cm Solid and Capsulated

Discussion

Spinal dermoid cysts are benign tumors located in the lumbar spine; the tumor cannot spread. However, the presence of these tumors can cause compression of the spinal nerves and at some point a cyst rupture can occur and the cyst will be resected.¹⁻⁴

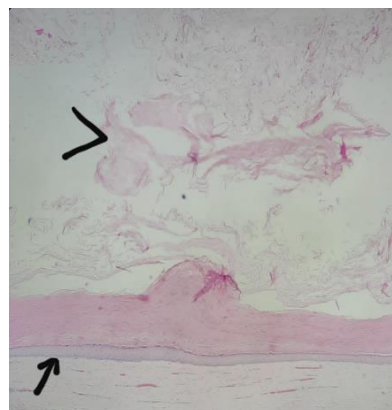


Figure 3 Microscopic View Show Cyst Lineage by Stratified Squamous Epithelium (Small Arrow) and Keratin Filled Lumen with Adnexa Structure (Large Arrow)

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A cyst is defined as a thin encapsulated structure that contains liquid secretion. Dermoid cysts contain components of the skin and are coated by a layer of skin that can remove dead skin cells. However, dead skin cells are not released into the environment out of the body, but will be collected in cysts. Therefore, the cyst will slowly enlarge. Dermoid cysts have skin cells, the production of components from these skin cells, other skin components (such as hair follicles, hair or sweat glands) and can even contain other components (such as components from teeth, oil, or blood).^{3,7}(Figure 3)

Previous studies revealed dermoid cyst intramedullary and intraspinal canal with a progressive neurological deficit before surgery.^{2,4-7} Dermoid cysts in the spine can be associated with other spinal abnormalities, such as spinal dysraphism, dermal sinus tract, spina bifida, spina bifida occulta, or myelomeningocele.⁷ Dermoid cysts can be discovered accidentally during treatment or diagnosis of one of these disorders.

This case is rare and unique because the dermoid cyst that occurred in extraspinal space causes compression of the surrounding soft tissue, no compression of neural structures. No transformation into malignancy during follow-up period. Spinal epidermoid cysts are uncommon. They comprise between 0.5% and 1% of all spinal tumors but account for up to 10% of intraspinal tumors in children.¹⁷⁻¹⁸ This makes it difficult to obtain data on case reports and epidemiology from the last five years.

Conclusion

Extraspinal dermoid cyst is a rare case need to be considered as a differential diagnosis for mass at sacrococcygeal. This condition requires special diagnostic and surgical modalities to provide better clinical results. Long-term survival with no complications could be achieved with complete resection. Early detection for cases like this is very important.

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