

## Case Report

## Squamous cell carcinoma arising from neglected meningocele

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**Abstract**

**Background:** A neural tube defect (NTD) is a common congenital anomaly with an incidence of 6.57–8.21 per 1000 live births. Patients usually present early because of obvious swelling or due to neurological deficit. However, neglecting the obvious cystic swelling on the back till its transformation into malignant tumor is rare.

**Case Description:** We describe a case of malignant transformation of meningocele in a 60-year-old man. Magnetic resonance imaging showed sacral meningocele. Neurological examination revealed intact motor and sensory examination with normal bladder and bowel function. There were no signs of meningitis and hydrocephalus. Excision was done and biopsy revealed it as squamous cell carcinoma.

**Conclusion:** Meningocele should be treated early and possibility of malignant change should be kept in mind in neglected cases presenting in adulthood.

**Key Words:** Adult, meningocele, neural tube defects, squamous cell carcinoma

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**Quick Response Code:****INTRODUCTION**

A neural tube defect (NTD) is a common congenital anomaly with an incidence of 6.57–8.21 per 1000 live births.<sup>[1,3]</sup> Patients usually present early because of obvious swelling or due to neurological deficit. However, neglecting the obvious cystic swelling on the back till adulthood is rare. To the best of our literature search, we could find only few such cases.<sup>[2,4-7]</sup>

**CASE REPORT**

A 60-year-old man presented with complaints of discharge from a swelling in the sacral area. At the time of birth he was noted to have a sacral meningocele for which he was advised surgery, however, his family had refused and the wound surface slowly became abraded and exudated repeatedly over a period of years. One month before the admission, the swelling started discharging foul smelling fluid and increased in size. Inspection showed a swelling in the sacral region of 5 cm in diameter, consisting of cauliflower-shaped swelling with yellowish slough

[Figure 1]. The area smelled foul and was constantly draining serosanguinous fluid. Neurological examination revealed intact motor and sensory examination with normal bladder and bowel function. There were no signs of meningitis and hydrocephalus. Magnetic resonance imaging (MRI) showed sacral meningocele with sinus tract [Figure 2a and b]. The tumor was excised, dural attachment was removed, and dura was closed again [Figure 3].

**Pathological finding**

The tissue sections were stained in hematoxylin and eosin (H and E) stain, and the histopathology study

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revealed tumor cells arranged in sheets and nests with keratin pearl formation [Figure 4], suggestive of well-differentiated squamous cell carcinoma. On high power examination, these tumor cells were large with high nuclei/cytoplasmic ratio and prominent nucleoli.

### Postoperative course

The postoperative recovery was uneventful and the wounds healed by primary intention. Further, the patient was sent to the oncology department for adjuvant therapy. Postoperatively, the patient has been on follow-up for a year without any recurrence.

## DISCUSSION

A meningocele is a congenital anomaly of neural arch fusion in association with an open neural tube defect, and is characterized by protrusion of spinal meninges which contain cerebrospinal fluid without involvement of the neural tissue. Most meningoceles are surgically repaired during the new-born period or at least in childhood. The incidence of survival is low without intervention,

and hence, adult meningoceles are rarely seen. Life expectancy at birth is shorter in myelomeningocele patients, although effective treatment for hydrocephalus and intermittent catheterization for the management of the neurogenic bladder can improve the quality of life for these patients. Posterior lumbosacral meningocele cases have rarely been reported. The long-term follow up results for adults with sacral myelomeningocele are not as good as in children because other neurological abnormalities such as hydromyelia, syringomyelia, tethered cord, Chiari malformations, and hydrocephalus accompany this lesion.<sup>[3]</sup>

A search of the literature revealed four cases similar to the one we have described. Saskun *et al.*<sup>[6]</sup> biopsied a neglected case of lumbosacral myelomeningocele who presented with fungating growth. Histology showed squamous cell carcinoma and radiotherapy was instituted. In the case reported by Thorp,<sup>[7]</sup> surgical treatment was chosen initially for a 26-year-old man with a carcinoma



Figure 1: Adult sacral meningocele with yellowish slough over it



Figure 3: Operative photograph showing the swelling being excised



Figure 2: (a, b) Magnetic resonance imaging of the spine (T1, T2 sagittal view) showing sacral meningocele

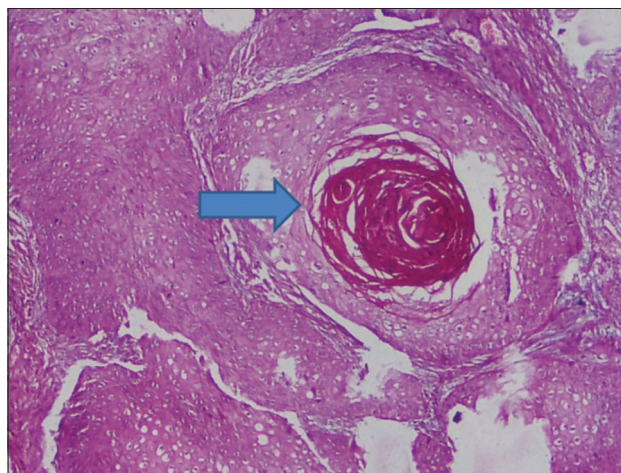


Figure 4: Photomicrograph shows tumor cells arranged in sheets and central keratin pearl (marked arrow), which is suggestive of it being squamous cell carcinoma (H and E stain, x400)

at the site of a lumbar meningomyelocele. Six months after resection, the tumor recurred and was treated with radiotherapy. Later, the tumor appeared again, necessitating further surgery. Three weeks postoperatively, the patient died of septicemia. In the case described by Pope and Todorovl,<sup>[5]</sup> a squamous cell carcinoma developed at the site of a cervical meningomyelocele in a 37-year-old man. This lesion was easily excised, without complications because the defect was a meningocele, and hence did not contain neural elements or connect with the spinal canal. Hong- Zhou *et al.*<sup>[2]</sup> excised a cauliflower-shaped lumbosacral myelomeningocele in an 11-year-old boy, who had the swelling since birth. It turned out to be squamous cell carcinoma on histology. Our patient presented as a neglected case of sacral meningocele with a discharge from the swelling and normal neurological examination. Excision of the mass was done and biopsy revealed squamous cell carcinoma.

MRI is a good investigation choice for evaluating the meningocele sac in the sagittal plane, and to observe the spinal cord itself, as well as the possible congenital anomalies associated with it.<sup>[2,4]</sup> In meningocele cases, neurological involvement are not seen as often as in myelomeningocele lesions, however, the local signs of sacral nerve involvement are seen as pain in both legs and bladder dysfunction. Taking this into consideration, somatosensory evoked potentials (SSEP) can be used in these patients as in myelomeningoceles.<sup>[4]</sup>

One must be aware of possible neoplastic change in untreated meningomyelocele after years of mechanical irritation and chronic bacterial infection. This seems to be a probable cause in our case. The defect, lacking a protective epithelial cover, must be viewed regularly with a high degree of suspicion, as with other chronic ulcers. Squamous cell carcinoma is a well-known complication

of burn and chronic venous ulcer, pilonidal sinuses, longstanding bacterial and fungal infections, vaccination scars, and even tattoos.<sup>[6]</sup>

Once change is noted, biopsies should be done to establish a histological diagnosis. If malignancy is established, an intensive search for metastases, lymph node involvement, and local invasion must be made for proper staging and subsequent treatment.<sup>[6]</sup>

## CONCLUSION

In conclusion, meningocele should be treated as soon as diagnosed and possibility of malignant change should be kept in mind in neglected cases presenting in adulthood.

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## Conflicts of interest

There are no conflicts of interest.

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