

# A rare cause of Sister Mary Joseph's nodule

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## Summary

Sister Mary Joseph's nodule is an important physical sign. It usually represents umbilical metastasis of intra abdominal malignancy. Its association with intra abdominal desmoplastic small round cell tumour is extremely rare. We describe a rare case of sister Mary Joseph's nodule associated with desmoplastic small round cell tumour. We

conclude that differential diagnosis of Sister Mary Joseph's nodule should include desmoplastic small round cell tumour, especially in adolescents and young adults.

*Key words: Sister Mary Joseph's nodule; desmoplastic small round cell tumour*

## Case report

A 28-year-old Caucasian male presented with a two-week history of episodic colicky central abdominal pain. The past medical history was significant for ulcerative colitis. On physical examination the abdomen was mildly distended but no mass was palpable. A violaceous periumbilical nodule (figure 1) was consistent with Mary Joseph's nodule, and bilateral firm, hard inguinal lymph nodes were present.

Abdominal and pelvic CT revealed a 5.5 cm lobulated mass lying between the anterior aspect of the rectum and bladder (figure 2, arrow), with several peritoneal deposits but no ascites. It also showed bilateral obstruction

of both ureters distally with bilateral hydronephrosis.

Histopathological evaluation of inguinal lymph node with small cell morphology. The tissue diagnosis of desmoplastic small round cell tumor (DSRCT) was confirmed on the basis of a characteristic t(11;12)(p13;q12) cytogenetic translocation.

The patient underwent bilateral nephrostomy. The neoplasm was inoperable and he is currently being treated by systemic chemotherapy.



**Figure 1**  
Sister Mary Joseph's nodule.



**Figure 2**  
Axial CT of abdomen and pelvis shows the mass in the rectovesical pouch (white arrow).

## Discussion

Sister Mary Joseph's nodule is a rare but important clinical sign. It usually represents an umbilical metastasis of an intraabdominal ma-

lignancy, with most of the lesions arising from the stomach, large bowel, pancreas and ovary. It is named after Sister Mary Joseph (1856–1939),

the superintendent of St Mary's Hospital in Rochester, who noted the association between umbilical nodules observed during skin preparation for surgery and metastatic intraabdominal cancer confirmed at operation [1]. Its association with intraabdominal DSRCT has rarely been described. To the best of our knowledge there have been only two previous reports in the literature of Sister Mary Joseph's nodule as a presenting sign of DSRCT [2, 3].

DSRCT is a rare, highly aggressive cancer usually affecting male adolescents and young adults. It was first described in 1989 by Gerlad and Rosai [4].

It is usually intraabdominal, often with diffuse peritoneal implants. Common presenting symptoms and signs are abdominal pain, palpable abdominal mass, abdominal distension, vomiting, hepatomegaly and palpable lymph nodes [5, 6]. Other associated features are intestinal and ureteral obstruction [6].

Histologically the tumour is characterised by well-defined nests of small round blue cells separated by abundant desmoplastic stroma. Cytogenetic study reveals a characteristic reciprocal chromosomal translocation, t(11;12)(p13;q12) [7].

CT findings include bulky intraabdominal soft-tissue masses involving omental and serosal surfaces, without a distinct organ of origin; solid, dominant, heterogeneous pelvic masses in the retrovesical or rectouterine spaces; and concurrent metastases common at the time of diagnosis, particularly those involving lymph nodes and liver [8].

Regardless of the treatment modality (chemotherapy, surgery or radiotherapy), the disease is almost uniformly fatal.

DSRCT should be considered in the differential diagnosis of Sister Mary Joseph's nodule, especially in a young male patient with an intraabdominal mass. Diagnosis is based on recognition of characteristic CT, histopathological and cytogenetic findings.

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