

## Sister Mary Joseph Nodule: Impact on the Clinician

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Once in a while in the dynamic world of dermatology practice, physicians will encounter a patient who will demand their fullest skills. A case of a patient presenting with a Sister Mary Joseph nodule (SMJN) caused me to reflect on how I responded to the diagnosis, how I dealt with the patient and his family, and how I have matured as a clinician.

An 86-year-old man presented with an asymptomatic umbilical lesion of a few months' duration. His medical history was remarkable for diabetes mellitus, hypothyroidism, mitral valve prolapse, and spinal stenosis. He was otherwise in excellent health and denied abdominal pain or weight loss. His primary care physician recommended that he see a dermatologist. Physical examination demonstrated a friable, focally eroded nodule in his umbilicus. Palpation of the lesion revealed a firm, fixed, deeper component. Because of the clinical suspicion of an SMJN, abdominal magnetic resonance imaging with gadolinium contrast was obtained, which revealed pancreatic cancer with metastases including the abdominal wall at the site of the umbilicus; these findings corresponded with an SMJN and evidence of biliary obstruction. While he was awaiting his scheduled appointment with the surgeon for a biopsy of the lesion, he developed a gastrointestinal bleed as well as obstructive jaundice, and he died 3 weeks following his dermatologic visit.

The medical aspect of this case is straightforward. The clinical suspicion of metastatic intra-abdominal cancer quickly was confirmed and the dismal prognosis associated with this sign unfortunately came to fruition quickly. However, what lingers is a reflection of our role as clinicians including how we handle our suspicions and concerns and how we assist a patient and his/her family during this most difficult time.

An SMJN is an umbilical nodule representing a metastasis from a malignancy that is characteristically of abdominal or pelvic origin.<sup>1</sup> Sister Mary Joseph Dempsey (1856-1939), a surgical assistant to Dr. William James Mayo, noted the presence of a hard umbilical nodule in a patient who was being prepared for surgery in 1928. Sir Hamilton Bailey coined the term *Sister Mary Joseph nodule* in 1949. The most common origins of SMJNs are gastrointestinal (52%),

gynecologic (28%), stomachic (23%), and ovarian (16%) carcinomas. Approximately 15% to 29% of all cases of SMJNs have an unknown origin and 3% of cases originate from the thoracic cavity. Primary tumors in several other sites including the gall bladder, uterus, liver, endometrium, small intestine, fallopian tube, appendix, cervix, penis, prostate, urinary bladder, breast, lung, and kidneys also have been reported to cause SMJNs.<sup>1</sup> Sister Mary Joseph nodules from pancreatic cancer are considered rare and account for approximately 7% to 9% of cases presenting with umbilical metastases.<sup>2</sup>

This case had a tremendous emotional impact on me. It forced me to assess my abilities as a physician, not just as a dermatologist. It was gratifying when the patient's widow called me to express her appreciation for my understanding and my help at that most trying time; she also commented that her late husband had been grateful. There were several important junctures in this encounter that required some decisions to be made rapidly. Obviously I made these decisions because I thought they were correct; perhaps others would handle certain situations differently. The purpose of this report is to have the reader consider how he/she would manage similar circumstances.

I was confident of the diagnosis. This type of lesion in an 86-year-old man was not going to be an omphalomesenteric duct cyst or urachal remnant. Interestingly, I had not previously seen an SMJN in my career (I began my dermatology residency in 1980). After examining the patient, several thoughts immediately crossed my mind: (1) How do I convey my concern, especially when the primary care physician told the patient and his wife it was a dermatologic problem? (2) How do I answer the inevitable question as to why the primary care physician did not reach the same conclusion? (3) How do I explain the portentous prognosis while offering some hope? (4) How can I help in any way; and (5) How can I do all these things when my office is crowded and I am already behind in my schedule?

The last question is the easiest to answer. I would take the necessary time to explain my concerns, knowing full well that for the rest of the day several patients would most likely complain of their wait. So be it. I cannot imagine conveying the gravity of the situation without allowing ample time for patients to comprehend what is being discussed. I instructed my front

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desk staff to tell patients that I would be running late. If they could not wait, their appointments should be rescheduled. I subsequently apologized for the delay to those patients who did not reschedule.

Although honesty is the best policy, I do not believe in brutal honesty. Even though I recognized that the patient presumably had widely metastatic disease with an ominous prognosis, I did not know that it was pancreatic in origin at that moment. I handled this situation by acknowledging my concern that the lesion was likely to be a malignant process, and if that proved to be the case, consultation with oncologists and surgeons would be necessary to determine the extent of the disease and formulate a treatment plan. Although the prognosis may be poor, I reminded him that there have been tremendous strides for certain tumors that were previously considered inoperable because of a focus on newer targeted therapies. We discussed the value of a second oncologic opinion and experimental protocols. Even in the direst circumstances, I think it is always appropriate to offer some hope. I explained to the patient that there are always choices to be made; he and his wife had to determine how to proceed. These decisions would even include choices related to palliative care.

As expected, the patient and his wife asked me why his primary care physician did not recognize this sign. Patients ask this question all the time; my usual response is that I can only explain what I am thinking and why. I have no doubt that one of my patients may go to another physician and ask what I was thinking. I explained to this patient that in more than 3 decades of practice, this was the first case that I have encountered with a (presumed) SMJN, which he should keep in perspective to explain why another physician may not have recognized it.

Inevitably, the patient wondered if he could have been diagnosed and treated earlier. I gently explained that once an SMJN is observed, the malignancy likely has already metastasized and therefore the ultimate prognosis may not be any different. This conversation was reiterated with his wife after he died. I told her that given his rapid progression, a delay in diagnosis by a couple of months may have been a blessing in that the prognosis may not have changed, yet they were enjoying life to the fullest during those months. Regardless, looking back was not going to help the patient. We needed to move ahead with diagnosing his condition. Because the patient and his wife were clearly paralyzed and devastated by this discussion, I needed to assist them by being somewhat assertive. I was able to

arrange for both surgical and oncologic evaluations and instruct them on where to go and who to see.

I first examined the patient on Monday; Friday afternoon I received a call from the radiologist confirming that the patient had (presumed) pancreatic cancer that had metastasized to the liver and abdominal wall. As promised, I called the patient and his wife as soon as I learned the results. I asked them if they would like to come to the office so we could discuss the diagnosis in person; they did not want to wait. We talked on the telephone for an hour, carefully going over the report. There were many questions, which I answered to the best of my ability. I had to remind them that I was a dermatologist, not an oncologist or surgeon. Much of the time was spent listening to their expressions of disbelief, fear, and anxiety. As the hour passed, I noted a sense of acceptance and a willingness to do what was necessary to battle this malignancy.

I called the patient and his wife the day after his appointment with the oncologist. They were delighted with him and his plan for confirming the diagnosis as well as the therapeutic plan. Unfortunately, his gastrointestinal bleed obviated that plan. I stopped by the intensive care unit to see him, now remarkably jaundiced and obtunded; we never spoke again.

It is easy to get caught up in the rapid-fire daily routine of the clinical and business aspects of medicine. Even the most devoted dermatologist can get sidetracked by headlines of cuts in Medicare reimbursement, pressure to generate relative value units, deciding whether to join accountable care organizations, navigating the transition to electronic medical records, and more. This patient presented with an SMJN due to metastatic pancreatic cancer and forced me to consider the fundamental essence of being a physician, which is to serve as a patient advocate. I also learned how I have changed over the years. As a medical student, I would have been enthralled to see this patient; now these cases sadden me. The challenges presented by this patient, however, garnered all of my skills as a physician and affirmed why this profession remains the most arduous and rewarding of all.

## REFERENCES

1. Dar IH, Kamili MA, Dar SH, et al. Sister Mary Joseph nodule—a case report with review of literature. *J Res Med Sci.* 2009;14:385-387.
2. Yendluri V, Centeno B, Springett GM. Pancreatic cancer presenting as a Sister Mary Joseph's nodule: case report and update of the literature. *Pancreas.* 2007;34:161-164.

### QUICK POLL QUESTION

All cases of a Sister Mary Joseph nodule are due to a primary intra-abdominal malignancy.

True

False

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