CANCER BOARD CONFERENCE

A case of retroperitoneal tumor with aortic aneurysm

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Abstract The differential diagnosis of retroperitoneal tumor associated with aortic aneurysm is a challenge. A 79-year-old frail male elderly, who had the history of lymphoma, was found to have a retroperitoneal tumor adjacent to the left common iliac artery aneurysm. Although there was no distant metastasis detected, the patient was in poor nutritional status, and he complained of pain in the lower left leg. The diagnostic procedure and potential treatment options were discussed in the cancer board. How to set the goal of his treatment was quite difficult. The patient decided to receive the best supportive care, and he did not undergo a biopsy of the tumor to make a pathological diagnosis.

Keywords Iliac artery aneurysm \cdot Retroperitoneal tumor \cdot IgG4-related disease \cdot Frail elderly \cdot Nutritional status

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Case presentation

Dr. Sasai (radiologist, conference chairperson)

Good evening, everybody. Welcome to the conference. Today's case is from the urology department.

Dr. Fukaya (urology resident)

The patient is a 79-year-old male. In April 2013, a local physician examined him for a left common iliac artery aneurysm, at which time a CT indicated left hydronephrosis. He was referred to the urology department at a regional hospital, where complete ureteral obstruction was found when a retrograde pyelogram was undertaken. Imaging was unable to yield visualization of the renal pelvis. The hospital concluded that obstruction was caused by the

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aneurysm. As the patient was 79 and had considerable thinning of the left renal parenchymal, renal function could not be restored, and the patient was kept under observation without treatment. In May 2013, the patient received stentassisted endovascular coil embolization in the left internal iliac artery to treat the aneurysm. There were no major postprocedural complications, but the patient continued to feel slight pain in the left lower back. The patient was subsequently treated for diabetes at our hospital and referred him to our department for treatment of a retroperitoneal tumor.

On January 4, 2014, a follow-up CT scan indicated progressive hydronephrosis and an enlargement of the retroperitoneal mass with a positive PET/CT. (Fig. 1) We considered a biopsy for a differential diagnosis of a tumor, infection, etc. However, the patient was a frail elderly in poor nutritional status and his weight had dropped from 70 to 55 kg during the preceding year. He suffered from anemia and fecal incontinence. However, he did not recall if he had previously gross hematuria. We therefore proceeded with a systemic evaluation and image studies. The patient has been discharged from the hospital and currently resides in a nursing home. We would like to benefit from your professional opinion regarding future treatment.

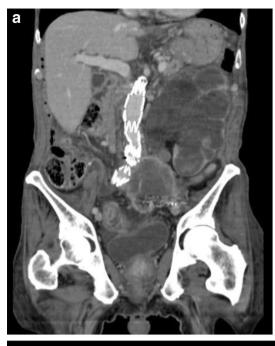
The patient is 79 years old who is now in a nursing home. He is confined to a wheelchair. His performance status is PS 2-3. The patient has no obvious signs of dementia. The left lower back pain is being treated with NSAIDs. The patient is unable to lie down for extended periods of time, and slept in a raised position during his hospitalization. A simplified nutritional assessment yielded a score of 16, signifying malnourishment. The patient's older sister lives nearby, but residing together with the patient is apparently not an option. There is no request for chemotherapy.

This CT image is from January of this year, at which time the retroperitoneal mass was approximately 75 mm in size. The mass is low grade with internal heterogeneity. We were unable to make a diagnosis.

The PET/CT seemed to point to an infected aortic aneurysm or abscess rather than a retroperitoneal tumor.

The CA19-9 tumor marker yielded a slightly high value of 100, with SCC also slightly high at 2.0. The PSA was 18.5 (Table 1).

The patient received treatment for a malignant lymphoma in the neck region in the past. It seems that the tumor was surgically removed and the patient healed following treatment, but this particular history dates back quite some time, and the details remain unclear. We therefore measured the IL2 receptor, but found it to be normal. If a differential diagnosis were to identify ureteral cancer, the TNM would likely be T4N0M0. In such a case, a complete cure would be difficult. The goal would instead be slowing the pace of progression or controlling



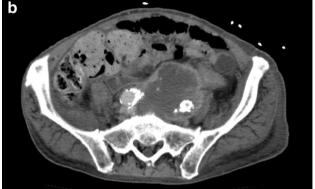


Fig. 1 CT scan showed that retroperitoneal mass was identified in the left iliac artery region that caused left hydronephrosis. **a** Coronal section, **b** transverse section

hemorrhaging through radiation therapy. One might also consider the possibility of a malignant soft tissue neoplasm originating in the retroperitoneum. Here is the CF finding of our examination of the colon. A neoplasm resembling a submucosal tumor was identified with the naked eye. We took a biopsy, and pathology identified an adenoma, but the biopsied sample did not allow us to rule out colorectal cancer. Diagnosis could point to another malignant lymphoma. If the diagnosis is ultimately colorectal cancer, we would like to hear your opinion as to whether curative treatment is possible, and whether radiation therapy would be appropriate.

Dr. Sasai

Thank you. Does anyone have questions concerning the medical history?

Table 1 Labo data

Tumor markers
CA19-9: 100 U/ml (0-37)
CEA: 2.4 ng/ml (0.0-3.0)
sIL2r: 452 U/ml (145-519)
NSE: 13.0 ng/ml (0.0-16.3)
Nutritional index
TP: 5.6 g/dl (6.5-8.5)
Alb: 2.5 g/dl (4.0-5.2)
Hb: 8.7 g/dl (13.4–17.1)
Ca: 8.1 mg/dl (8.8-10.6)
Ca: 8.1 mg/dl (8.8–10.6)

Dr. Kato (medical oncologist)

When was the lymphoma treated?

Dr. Shiosawa (urologist)

Approximately 15-16 years ago.

Dr. Sasai

And the tumor was surgically removed with no subsequent chemotherapy?

Dr. Fukaya

Yes, that's correct.

Dr. Horikoshi (general medicine)

How about the histopathology?

Dr. Shiosawa

Diffuse large.

Dr. Kato

If it were a DLBCL, wouldn't there have been pathological change in other organs? It would be hard to imagine not administering chemotherapy. If chemotherapy was not administered, there must have been a good reason. Considering the aggressive lymphoma, one cannot rule out a recurrence, but lymphoma seems unlikely in this particular case. In terms of diagnosis, the only biopsy specimen came from the colon, right?

Dr. Fukaya

Yes, that's right.

Dr. Sasai

All right, then, Dr. Suzuki—would you kindly give us your opinion based on the imaging?

Dr. Suzuki (diagnostic radiologist)

As of April 2013, the image shows an adherent mass to the iliac artery aneurysm. It increases in size over time (switching to the stent placement CT), explaining, I think, the hydronephrosis. That is basically what I can contribute from the imaging.

Dr. Sasai

Moving on, then, the retroperitoneal mass was identified in the iliac artery region in April 2013. It gradually increased in size, and an endoscopic biopsy was performed to determine whether it had invaded the sigmoid colon, or whether the sigmoid colon had alternatively undergone some pathological change of its own. However, the biopsy did not yield any finding of malignancy. So if it is not ureteral cancer, what else could it be?

Dr. Fukaya

A submucosal tumor or a neuroendocrine tumor, perhaps.

Dr. Sasai

Let's discuss the possibility that this patient may have colorectal cancer.

Dr. Sakamoto (colorectal surgeon)

I think the chronology negates that possibility. However, even if it were possible, it would be T4, surrounding the great vessels, and could not be surgically removed.

Dr. Suzuki

The imaging seems to suggest it could be an infected aneurysm, but does the lab data show any change? A determination can only be made through tissue pathology.

Dr. Shiosawa

CRP was 2.0, with no clear evidence of a high WBC or elevated procalcitonin or other marker for septicemia. I find it difficult to believe that this is an abscess.

Dr. Sasai

What about the pathology? Can we conclude this is a straightforward adenoma?

Dr. Arakawa (pathologist)

We see no change in the neoplasm from mucous membrane hyperplasia, and urine cytology with paracentesis yielded a class IIIb finding, which was insufficient to sustain a cancer diagnosis.

Dr. Sasai

...so the pathology is also inconclusive, then.

Dr. Kato

What about the site from which the biopsy specimen was harvested? The CF image seemed to leave the possibility of cancer open.

Dr. Fukaya

Biopsy specimens were taken at multiple sites, with a reasonable distance between each.

Dr. Sakamoto

I don't think we see any particular change in the colon to indicate cancer from the CF image. The biopsy site also looks fairly normal to me.

Dr. Sasai

This case presents a real diagnostic challenge. Does anyone have any further input?

Dr. Horikoshi

Just to reconfirm ... could you go over the tumor markers once again?

Dr. Fukaya

The CA19-9 was about 100, and the SCC was slightly elevated at 2.0.

Dr. Horikoshi

... and the CEA?

The CEA was normal. 2.4.

Dr. Sasai

Right. So how do we proceed with treatment?

Dr. Horikoshi

We hear a lot about IgG4-related disease these days. Were you able to look into that?

Dr. Shiosawa

There is no particular indication of that in this case.

Dr. Horikoshi

I think we had to better make a differential diagnosis of IgG4-related disease.

Dr. Shiosawa

IgG4-related disease of course indicates inflammatory disease, but in the absence of clinical findings, doesn't the possibility remain low?

Dr. Sasai

What about the possibility of ureteral cancer?

Dr. Shiosawa

We initially planned on a biopsy, but in consideration of the patient's poor nutritional status, we opted to pursue diagnosis through minimally invasive procedures and therefore hospitalized the patient for a systemic evaluation. We began considering options, and that has brought us to where we are today.

Dr. Sasai

How about CT-guided biopsy?

Dr. Shiraishi (diagnostic radiology)

CT-guided biopsy might be feasible. Imaging might even allow for a posterior approach.

Dr. Sasai

Returning to the question of treatment, then—assuming ureteral cancer, what treatment would the urology department likely pursue?

Dr. Shiosawa

Assuming T4, we would likely administer chemotherapy or pursue palliative radiation options. Considering the patient's overall condition, aggressive chemotherapy would be unlikely.

Dr. Sasai

Now a question for colorectal surgery... Since the patient is under observation at the moment, what about the likelihood of a future colostomy?

Dr. Sakamoto

Quite likely, I think, as a palliative measure.

Dr. Sasai

What measures would the radiation team consider?

Dr. Ishikura (radiation oncologist)

This is a challenging case. The problem is deciding on the objective of the palliative measure. Considering the size and site of the mass, radiation therapy might not help. Does the patient's condition even allow for consideration of palliative radiation therapy?

Dr. Shiosawa

Due to lower limb pain, the patient is unable to lie down for any sustained period. The tumor is probably compromising the iliopsoas muscle; the patient is receiving oral medication for relief and shows an improved PS. Paracentesis of the renal pelvis yielded a finding of dark red blood in the urine. As the patient has complete blockage of the ureter, this is not a problem, but if hemorrhaging becomes an issue, I think radiation therapy might be appropriate. Does anyone have any thoughts on this?

Dr. Sasai

That is based on the presumption of ureteral cancer, isn't it?

Dr. Shiosawa

Exactly. If the tumor invades muscle near the great vessels, is it correct to assume that radiation would be virtually impossible?

Dr. Sasai

There would be danger of perforation and hemorrhaging, I would think.

Dr. Shiosawa

If the mass is in fact benign, left renal function is almost fully compromised at this point, but there is no current danger of infection, so continued observation should be possible.

Dr. Sasai

The patient is not experiencing pain at present, as I understand. Do we therefore conclude that this is not the time for aggressive interventional therapy?

Dr. Horikoshi

Would QOL diminish significantly if the patient began to experience pain?

Dr. Shiosawa

That would likely depend on the degree of invasion into muscle, but the patient's mobility would surely be impacted.

Dr. Takagi (orthopedic surgeon)

I think continued observation would be appropriate as long as symptoms are controlled by medication. However, if the patient were to lose mobility due to stiffening legs, a different course of action would need to be addressed.

Dr. Sasai

What about the nursing care perspective? The patient is currently in a nursing home, I believe.

Dr. Nakano (oncological nurse specialist)

I am not familiar with the specifics of the patient's condition, but reevaluating for dementia and other lifestyleimpacting factors is necessary, I would think.

Dr. Sasai

Very well. Let me wrap up what we have covered.

A tumor of unknown etiology appeared in the retroperitoneum. The patient has a history of treatment for lymphoma. The patient currently has pain in the lower left leg and has restricted mobility, but the symptoms are being controlled with oral pain medication. The patient is in a nursing home. The patient's wishes, and those of his family, are unknown, but the cancer board recommends continued observation, if I understand correctly. Symptomatic treatment of pain remains under discussion, and a colostomy may be considered in the future if gastrointestinal obstruction becomes an issue.

Are there any additions or corrections?

Dr. Sasai

Thank you for your participation. Quite many of you made time for this meeting today. I think we had a lively discussion about this challenging diagnostic case. Thank you all.

Follow-up

Horie (Urologist)

The multidisciplinary discussion concluded that ureteral cancer was the likely diagnosis of this retroperitoneum tumor. Colon cancer and the recurrent lymphoma, IgG4-related disease was probably unlikely. CT-guided biopsy of tumor was recommended as a less-invasive diagnostic measure.

We communicated the content of the cancer board discussion to the patient and his older sister who accepted that diagnosis would be invasive and that the patient's condition could not easily sustain the ongoing treatment. The patient and his family did not desire to pursue the pathological diagnosis by a biopsy of the tumor. Future treatments will prioritize alleviation of symptoms to help sustain the patient's lifestyle. The patient is currently in a nursing home, and the attending physician sees him weekly for palliative care. His pain is being controlled by NSAIDs, and the patient is receiving lymphatic massages on his lower left thigh.

Nutritional status was not discussed in this cancer board. Several studies have demonstrated the importance of weight loss as a prognostic factor for survival in patients with cancer. Presumably, the weight loss of this patient was due to the inflammatory response by the retroperitoneal tumor. However, we might have introduced screening tool for nutritional status such as the Mini-Nutritional Assessment [1]. Moreover, poor nutritional status was associated with a higher degree of depressive symptoms as well [2]. A comprehensive geriatric assessment (CGA) includes an evaluation of an older individual's functional status, comorbid medical conditions, cognition, nutritional status, psychological state, and social support, and a review of the patient's medications [3]. The use of CGA would be useful to decide the treatment options and improve the care of older adults with cancer.

Conflict of interest The authors declare that they have no conflict of interest.

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