Personal Paper

THE OPHELIA SYNDROME: MEMORY LOSS IN HODGKIN'S DISEASE

IAN CARR
Department of Pathology, University of Saskatchewan, Saskatoon, Saskatchewan S7N 0W0, Canada

I HAD sat by the fireside a month or two previously and said to Jane, who was 15½—"You're the only one of the bunch who has never caused me a moment's worry". It was true. Some kids seem to dance through childhood, into the teens, on lightsome feet, proof alike of God's grace and the wayward benevolence of genetics. She had come from a village in middle England to a small Canadian university city three years previously. She didn't greatly like it but made the best of it. The young Canadian kids she brought home looked a bit sloppy at first. But we learned that her taste in friends was unerring and that Canadian and English teenagers differed only on the surface.

Jane had been near the top of her class. She had "glandular fever" and did a little less well—but the light still danced in her green eyes. Then, when we walked the dog one evening, something seemed different. I had often read to her the old Scots ballad of Bonnie Kilmeny—the girl who was spirited off by fairies. A little of the sparkling precision of her conversation had gone. But if you have four kids you can't worry all the time.

I was away when it happened. She forgot to let a friend's dog out over the weekend and lost the memory of that weekend. Over a beer a paediatrician friend who had seen her because of amenorrhoea said he was worried—he didn't know why. A good neurologist could find nothing wrong. But Jane couldn't remember.

We went on holiday. All would be well—nothing would be wrong with Jane. Super Jane we called her. She had kept us right in the early dizzy days in our new country and organised well my early dotage. Now we lay on the sand of Vancouver Island. Jane and I swam. We walked and visited the small islands; and she was her quaint, cool, remote self, with the infinite promise of femininity lurking around the corner of her smile. Or was she perhaps more remote, and a little hazy about what we did yesterday? Nothing could be wrong with Super Jane, who had ridden on my shoulder the day before yesterday.

Home, and back to work; grade 11 was hard. Super Jane would manage—and off she went at 7.30 A.M., her lissom confident self. But there was never any homework; Super Jane did it at school. It took a month before a conversation with a school teacher made me wonder, and ask to see some homework.

A golden summer crisped into fall. Jane and I sat night by night, with homework. If she brought it home, if she brought the right books, if she brought any books. By the middle of October we knew there was something seriously wrong. We realised when we looked at her work and talked to worried teachers that there was a serious problem – a teenage neurosis, perhaps drugs. The evening walk became more bizarre, like talking to an elderly person whose conversation slides into a twilight of fading concern and randomly illuminated glimpses of the past. She was now aware that there was something wrong and that she, the intellectual, was somehow less effective at school than many of her previously despised "dumb" classmates.

No friends now came. The telephone calls, the evenings out, of a blooming adolescence—all gone. Her life closed down to rising in the morning for school, as always on time; a strange secluded time at school, of which she didn't talk; and then back to home and parents. We were half cross with her "idleness" and half worried.

Our friend and family doctor of course had seen her; and a check on her endocrine status had been run by an endocrinologist. There was nothing abnormal. At last a psychiatric referral. My mind, which had been talking to Jane through a glass wall for weeks, leapt to the hebephrenic schizophrenia of the psychiatry texts of twenty years ago. I remembered Ophelia. Yet there was something odd. At this point, I began to wonder, still immigrant, was our small Canadian medical school good enough for Jane, or would we fly her back to England? I was to learn.

The psychiatrist was helpful, kind, and in the event one of the best doctors I have ever met. He was unwilling to commit himself to a name for this psychiatric syndrome. She was depressed; he thought depression needed treatment and wisely treated it with appropriate drugs. Two weeks later her feet started to sink into the carpet, and the floor to lift. Hallucinations. She was in hospital early next morning. We knew now that Super Jane would never be quite the same—nor would we. She was interviewed, talked to, and tested psychometrically, and (by now her blood cobalt and urinary rhubarb had been measured at least three times) as a standard precaution, her chest was X-rayed. There was a considerable mass in one lung.

The doctors were kind and talked of sarcoid. But it was clear to me that Jane had some sort of paraneoplastic syndrome. Jane herself was now aware of something wrong and yet somehow not too worried. In the next weeks Jane went from hallucinations into a parkinsonian state under the influence of the necessary drugs. A grey shaking shadow sitting in a corner of the room asking in a faraway terror-stricken voice, "What is happening to me? Why me?"

Mediastinoscopy clinched the diagnosis, and the sections were the saddest that ever crossed my microscope stage. Hodgkin's disease. In a psychotic. What right did we have to leave a psychotic in a twilight world for God knew how long? Jane, as she had been, would not have wanted it. There was considerable published evidence that the changes might, if related to the cancer, be due to an irreversible demyelinating lesion or a neurotropic virus. Could we allow staging of the Hodgkin's disease and treatment? The psychiatrist said we must not lose hope—and the medical machine ground on. Laparotomy. Splenectomy. Could an apparently psychotic frail teenager stand it? She did. Her psychotic condition got

DR RODERICK SMITH AND OTHERS: REFERENCES—*continued*

3. Aspin J, Sheldon M. An epidemic of tuberculosis in a Staffordshire school. *Tubercle* 1965; **46:** 321–44.
4. Rao VR, Joanes RF, Kilbane P, Galbraith NS. Outbreak of tuberculosis after minimal exposure to infection. *Br Med J* 1980; **281:** 187–89.
5. Masashi Tamura, Ogawa G, Sagawa I, Amano S. Observation on an epidemic of cutaneous and lymphatic tuberculosis which followed the use of anti-typhoid vaccine. *Am Rev Resp Dis* 1955; **71:** 465–72.
6. Heycock JB, Noble TC. Four cases of syringe-transmitted tuberculosis. *Tubercle* 1961; **42:** 25–27.
7. Stewart CJ. Tuberculous infection in a paediatric department. *Br Med J* 1976; i: 30–32.
8. Loudon RG, Roberts RM. Droplet expulsion from the respiratory tract. *Am Rev Resp Dis* 1967; **95:** 435–42.
9. Department of Education and Science Protection of school-children against tuberculosis. 1969, Circular 3/69
10. Department of Education and Science. Control of tuberculosis. 1978, Circular 12/78.

worse–and all through this time, despite the anguish of dealing with someone who was mentally very sick, and in a ward with some even sicker people, her friends still came, week by week, to see her and sit and talk to her.

Two courses of chemotherapy (we felt) should do something if chemotherapy was going to work. Intravenous drugs. Bruised arms. Vomiting. Falling hair. We had been in to see her every day for three months and now went away for a weekend. When we came back she remembered what she had been doing the day before. Not just a straw in the wind but bending branches. It went on. More chemotherapy. The snow went and the instant prairie summer came. Day by day she was remembering more and more. Radiotherapy–more vomiting. The whole back of Jane's head bald. More radiotherapy. The oddest choice I have had to make; what do you give your teenage daughter the day she starts radiotherapy? Flowers of course. At last a weak, wan Jane–the back of her head bald–went on holiday. Soon Jane was remembering.

She struggled back to normality, her mind returning to normal as her hair did. A year later the only apparent mental abnormality was a neatly excised piece of memory for about eighteen months. The factual information has been relearned, the personal development reundertaken. Only a bit of life is missing.

We have learned about a rare neurological phenomenon. The limbic syndrome–loss of memory for recent events–is rare in any case; it may be due to metastasis or demyelination associated sometimes with papovavirus infection and is usually irreversible, but there are a few instances where dialysis relieves the condition, implying that there is perhaps a circulating neurotransmitter-like molecule produced by the neoplasm. The usual cause of this rare condition is bronchial carcinoma. Hodgkin's disease is a very rare cause indeed. A computer search of the literature did not find a similar case. The neurological effects of Hodgkin's disease are reviewed by Kaplan.[1]

We learned that the standards of care in our university hospital are extremely high and are matched by the humanity of my colleagues. That the courage of a 16-year-old girl can meet extreme demands. That most of the concerns that beset the tranquillity of our daily lives are as chaff before the wind when it really blows; and it bloweth where it listeth. That our children, like our possessions and our lives, are lent not given. As Jane is lent, once more.

I record this story because it may help someone to a difficult diagnosis. In summary, recent memory loss may rarely be due to Hodgkin's disease, probably as a paraneoplastic event. It may be reversible and can be remembered as the Ophelia syndrome.

Our debt to many medical and nursing colleagues in University Hospital, Saskatoon, and Saskatoon Cancer Clinic is very deep indeed.

Correspondence should be addressed to I.C., Department of Laboratory Medicine, St Boniface General Hospital, 409 Tache Avenue, Winnipeg, Manitoba R2H 2A6, Canada.

REFERENCES

1. Kaplan H. Hodgkin's disease. Cambridge, Massachusetts, and London: Harvard University Press, 1980.

Disabilities and How to Live with Them

HAEMOPHILIA

I AM a 36-year-old haemophiliac with a coagulation factor level of less than 1%. Diagnosis was established when I was 2 years old after I had fallen and cut my forehead. Haemophilia was nothing new to my mother. Two of her brothers, now deceased, were severe haemophiliacs. She had discussed with her doctor the possibility of transmitting the disease to her children. His advice was that such transmission was most unlikely since haemophilia usually misses a generation.

My childhood bleeding was primarily to knee, elbow, and ankle joints, with nosebleeds and bleeding from superficial cuts and abrasions. I could always recognise a bleed by a bubbling, a tingling, or simply an awareness that all was not well around the area concerned. Each joint bleed followed the same depressing, predictable sequence of events: the early warning sign would be followed over a 3–6 hour period by a gradual loss of movement and an increase in pain and discomfort. My mother was usually the first to be told of a bleed and accepted the news in her usual calm, matter-of-fact way. This reassurance was of the utmost importance as it helped me to accept and cope with each incident. The reaction of my father was always of surprise and disappointment for me that I was again out of action, but he would quickly accept the situation and, like my mother, get down to giving me the care and attention I needed. My parents and I soon found that the local hospital had little understanding of the disease and was unable to provide any treatment. The local general practitioner was similarly out of his depth and was able only to advise rest. As our experience increased, we realised that we were the only ones who could decide what to do each time. Each bleed prompted the familiar question: "How did it happen?" Most of the time, it was impossible to remember. We did not appreciate that with such a low clotting level, trivial incidents could trigger off a bleed and bleeding could also occur spontaneously.

For bleeds in joints it was necessary to immobilise the limb completely until swelling and pain had subsided. Each episode took weeks and even months before full movement was regained. As the frequency of bleeds into knee joints increased, there was a gradual loss of full bending movement. I particularly dreaded bleeds into the knee or ankle joints as either rendered me almost helpless. My parents' lives were constantly disrupted with one or other having to stay with me during the night. Hot or cold compresses were applied, pain killers given, and anything that might ease the continuous agonising pain was done. Recovery was always slow and my days were spent propped on a sofa or bed, filled with boredom, loneliness, and an intense feeling of isolation from the outside world. Friendships with other children were difficult because of prolonged absences. When I was with them I was always torn between the irresistible attraction of joining in the more hazardous pastimes and the warnings from my parents.

Primary education proved impossible. Frequent bleeds resulted in poor attendance at school and after two years my education ceased. It was not until I was 12 years old that the education authority arranged for me to have home tuition. During those few years I reached a reasonably high standard of education. I later attended two colleges where my training was completed for future employment. At the age of 15 I had teeth extracted for the first time. Prior to admission to the local hospital, gum shield impressions were made of both sets of teeth. Immediately after the extractions the shields were firmly inserted, a method which fortunately proved successful, since there was still no known treatment. As I entered adolescence, the number of bleeds into joints began to